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### **Children Asthma Differential Diagnosis**

## Mohammad Gharagozlou <sup>1</sup> © P

Abstract: Children asthma is a multifactorial and heterogeneous disease with different phenotypes. This variety of clinical manifestations is caused by genetic variation and complexity. The primary goal of treating children with asthma is to control the symptoms of the disease so that the chance of acute asthma exacerbations and the progressive decrease in lung function can be significantly reduced. When the asthma disease does not respond well to the expected maintenance treatment, it is necessary to re-evaluate the initial diagnosis of the disease and in this regard the differential diagnoses of asthma such as immunodeficiency diseases, cystic fibrosis, primary ciliary dyskinesia, vocal cord dysfunction, anatomical abnormalities and vascular disorders of airways should be investigated, that the importance of each is different according to the age of the patient. If the diagnosis of asthma is definite, it is necessary to perform a more detailed assessment regarding the lack of appropriate response to treatment (difficult-to-control). In this context, the important and common comorbidities of asthma such as gastroesophageal reflux, sinusitis, rhinitis, obesity, anxiety and depression, and vocal cord dysfunction should be investigated. It is always necessary to consider common and correctable reasons for lack of proper response to treatment, such as patient and family nonadherence to prescribed treatments, poor drugs inhalation technique, the presence of allergens and polluting irritants in the patient's living environment, and psychological factors. Considering the complexity of diagnosis and treatment in children suspected of having asthma, or those with confirmed diagnosis asthma without proper response to treatment, it is necessary to take advantage of the consultation with allergists and pulmonologists for the purpose of multidisciplinary patient management.





### Immunology & Allergy

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### chronic asthma management

آرش كلانترى  $^1$   $^{\circ}$ 

Abstract: Asthma is a chronic intermittent obstructive lung disease affecting >230 million people worldwide and a significant cause of morbidity in patients of all ages. In many patients, asthma disease can be controlled with appropriate medical therapy. Even for many of the more severe asthma patients, significant advances in medical care have reduced complications such as exacerbations and also improved quality of life. The goal of asthma treatment is symptom control and prevention of future exacerbations. A stepwise approach to pharmacologic treatment is recommended The initial choice of medication is determined by the asthma severity classification. A step-up or step-down therapy is recommended depending on symptom control based on GINA guidelines Complete control of asthma is defined as: 1- no daytime symptoms ,2- no night-time awakening due to asthma ,3- no need for rescue medication ,4- no asthma attacks ,5- no limitations on activity including exercise normal lung function ,6- in practical terms FEV1 and/or PEF>80% predicted or best 7- minimal side effects from medication After starting or adjusting medicines for asthma, review the response to treatment in 4 to 8 weeks In general, people with asthma should use the smallest doses of ICS that provide optimal control for their asthma, to reduce the risk of side effects.





### Laboratory tests in Antiphoshpholipid syndrome

نيلوفر شعشعاني  $^1$   $^{\odot}$   $^{\odot}$  نيلوفر شعشعاني  $^2$ 

دانشیار، روماتولوژیست کودکان ۱ فلوی روماتولوژی کودکان دانشگاه علوم پزشکی بهشتی <sup>۲</sup>

**Abstract**: Laboratory tests in Antiphospholipid syndrome Mehrnoush Hassas Yeganeh, MD Niloofar shashaani, MD The diagnosis of antiphospholipid syndrome (APS) relies on the detection of antiphospholipid antibodies (aPL). The aPL play a crucial role in the diagnosis of APS. However, the laboratory diagnosis of APS remains challenging. Progress has been made to address some of the methodological challenges of all tests currently available, but an international reference material to overcome the inter-assay and inter-laboratory variation is still lacking. To obtain optimal performance, all assays have to be performed according to the guidelines. Currently, lupus anticoagulant (LA), anticardiolipin (aCL), and antibeta2glycoprotein I antibodies (a\beta 2GPI) IgG or IgM are included as laboratory criteria, if persistently present. LAC measurement remains a complicated procedure with many pitfalls and interfered by anticoagulant therapy. Solid-phase assays for aCL and aβ2GPI show interassay differences. These methodological issues make the laboratory diagnosis of APS challenging. In the interpretation of aPL results, antibody profiles help in identifying patients at risk. Other aPL, such as antibodies against the domain I of beta2-glycoprotein (aDI) and antiphosphatidylserine-prothrombin (aPS/PT) antibodies have been studied in the last years and may be useful in risk stratification of APS patients. Because of the methodological shortcomings of immunological and clotting assays, these non-criteria aPL may be useful in patients with incomplete antibody profiles to confirm or exclude the increased risk profile. All three assays, LA, \(\beta\)2GPI-dependent aCL, a\(\beta\)2GPI IgG, and IgM should be performed at the same time to increase diagnostic utility, with an integrated interpretation of all results and an interpretative comment. Making antibody profiles including LA, aCL and aβ2GPI help identify patients at risk. Confirmation of a positive result after 12 weeks is required since only persistently positive results are clinically relevant in the context of APS. Results should be interpreted in a clinical context and





### Immunology & Allergy

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### Management of food allergy

### Alireza Shafiei <sup>1</sup> <sup>®</sup>, Masoud Movahedi <sup>1</sup> <sup>®</sup>

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**Abstract**: Adverse food reactions have a high prevalence of about 20%. Food intolerance is an adverse reaction to foods due to factors inherent in the consumed food, such as toxic contaminants. Food allergy is an adverse reaction to foods caused by the immune system. Food allergy management includes avoidance of that food and proper and quick treatment of allergic reaction in case of accidental consumption of that food. Avoidance of food allergens should be based on a confirmed diagnosis. Despite the existence of diagnostic tests for the diagnosis of food allergy, these tests have limitations, and the oral food challenge (OFC) test is an essential part of the diagnosis and management of food allergy. Recommendations regarding food avoidance should be based on the specific condition of each patient and the severity of the hypersensitivity reaction. Avoiding food allergens involves educating the patient on how to prepare safe foods at home, reading food labels, and avoiding allergens in restaurants and schools. In order to prepare food, it should be kept in mind that the tools that are used to prepare or serve food and have been in contact with allergens should not be used, unless they are completely washed first. Patients with food allergies should be taught that non-food items such as medications, vaccines, cosmetics, etc., may contain food. Careful planning for patients with food allergies should be done to ensure that the nutrients in the eliminated food(s) are adequately replaced with appropriate food sources. Due to the possibility of unintentional or accidental ingestion of food allergens, patients should be taught to treat unexpected reactions after consuming food allergens in any situation. Allergen-specific immunotherapy by oral, sublingual and subcutaneous methods, alone or in combination with anti-IgE monoclonal antibodies and probiotics, are new treatments under investigation for the management of food allergy.





### THE DIAGNOSTIC APPROACH TO FOOD ALLERGY

منصوره شریعت  $^1$   $^{ ext{ }}$ 

Abstract: THE DIAGNOSTIC APPROACH TO FOOD ALLERGY In the past two decades, food allergy is an important public health problem. Food allergies are most common in childhood. Food allergy is suspected when typical symptoms (urticaria, edema, wheezing, mouth itch, cough, nausea, vomiting, anaphylaxis) occur within minutes to hours of food ingestion. A complete history of the reaction to each incriminated food is essential for proper diagnosis. Food allergy is defined as "an adverse reaction happening from a specific immune response that occurs reproducibly on exposure to a given food." Immunoglobulin E (IgE)mediated food allergies, or hypersensitivities, occur frequently in young children and account for most food-allergic disorders, although several non-IgE-mediated immune reactions, have been recognized. The diagnostic approach to food allergy begins with the medical history and physical examination. These assessments guide the selection of the laboratory tests. but history alone should never be used to make a diagnosis. Skin-prick tests are used to screen patients with suspected IgE-mediated food allergies. Food extracts are applied by the prick or puncture technique. Until then, a positive skin-prick test should be explained as indicating the possibility that the patient has symptomatic reactivity to the specific food, whereas negative skin test results confirm the absence of IgE-mediated reactions. In vitro allergen-specific IgE tests are used for measuring serum for IgE-mediated food allergies. Oral food challenges are an important in the management of food allergies in patients, and they remain the most accurate tests for the diagnosis of food allergy. Oral food challenges have significant risk, but these risks can be minimized by appropriate dosing and by performing challenges in a controlled setting with expert personnel.





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### Primary Antiphospholipid syndrome



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**Abstract**: Antiphospholipid syndrome (APS) is characterized by venous or arterial thrombosis and/or an adverse pregnancy outcome in the presence of persistent laboratory evidence of antiphospholipid antibodies (aPL). APS occurs either as a primary condition or in the setting of an underlying disease, usually systemic lupus erythematosus (SLE). APS can be further classified according to the type of clinical manifestation: thrombotic or obstetric, in some cases, both may be present and whether there is life-threatening multiorgan involvement. Thrombotic APS is used to describe patients diagnosed with APS based on venous or arterial thrombosis and persistent laboratory criteria for aPL. Obstetric APS is used to describe patients diagnosed with APS based on an APS-defining pregnancy morbidity (including fetal death after 10 weeks gestation, premature birth due to severe preeclampsia or placental insufficiency, or multiple embryonic losses [before 10 weeks gestation]) and persistent laboratory criteria for aPL. Catastrophic APS (CAPS) is a rare, severe (life-threatening) form of APS characterized by thrombotic complications, usually microvascular, affecting multiple organs that develop simultaneously or over a short period of time. Implications of these sub-classifications for management are discussed separately. Antiphospholipid antibodies – aPL are a laboratory finding. When persistent, they are a component of the clinical syndrome of APS. They can also be seen as a transient finding following infection or other acute illness. DIAGNOSTIC EVALUATION — Medical history, physical examination, and laboratory testing for antiphospholipid antibodies.





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# Antiphospholipid Syndrome (secondary to infections and other diseases)

shima salehi <sup>1</sup> © P

Abstract: Antiphospholipid Syndrome (secondary to infections and other diseases) The antiphospholipid syndrome (APS) is a prothrombotic disorder with recurrent venous and arterial thromboses and/or pregnancy morbidity in association with the presence of antiphospholipid antibodies (aPL) and defined by the presence of at least one clinical and one laboratory criterion. Patients with primary APS have no other autoimmune conditions, whereas secondary APS is diagnosed where the criteria for APS are fulfilled in the presence of another condition – most commonly systemic lupus erythematosus (SLE). APL has been reported in patients with: • Autoimmune rheumatic diseases • Infections • Malignancies • In association with certain drugs But these aPL only seem to cause true APS in association with autoimmune rheumatic diseases and, rarely, with some infections. Prevalence of aPL antibodies in children without any underlying disorder are generally higher than those reported for adults, may be related to the frequent occurrence of infectious processes during childhood. Anti-CL antibodies detected in APS are thrombogenic and "β2GPI dependent", but in patients with infections are "\$2GP independent". Some reports showed the association of aCL with vascular events in patients with a variety of malignant conditions, including solid tumors and lymphoproliferative malignancies. Several drugs may induce generation of aPL with a low prevalence and no clear association with APS clinical manifestations. The percentage of progression to SLE or lupuslike disease in pediatric patients with PAPS is double compared with that found in the adult PAPS patients. The majority of the evidences suggest that patients with PAPS and SLEassociated APS have similar clinical and laboratory profiles, with some exceptions. Although aPL antibodies have been studied most extensively in association with SLE, they have also been reported in patients with other autoimmune disorders (systemic sclerosis, polymyositis/dermatomyositis, early undifferentiated connective tissue diseases, Sjögren, ANCA-related renal vasculitis).





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### Antiphospholipid syndrome treatment in pediatrics

### Raheleh Assari <sup>1</sup> © P

Abstract: Antiphospholipid syndrome treatment in pediatrics Abstract: In children, the treatment of antiphospholipid syndrome is in a challenge in comparison to adult; The lack of validated diagnostic criteria, no valuable clinical trial study or prospective study because of rarity in children and concern about the complications of treatment such as bleeding with notice of more activity in children and low adherence to treatment. Therefore, the managements of antiphospholipid in children are according to adult guidelines.1, 2 There are no definite treatment for APS to cure or eliminate the APL. The goal of treatment is to reduce the blood clot formation, blood thinner considering prevent the complications. The mainstay of treatment is to keeping the blood thinners after blood clod in patient with antiphospholipid positive. Aspirin, Warfarin, low molecular- weight heparin are used to prevent the clot formation. Blood clot is initially treatment with heparin followed by long term warfarin. In children with aniphospholipid positive without clot the treatment outweigh the unproven benefit of aspirin with complication of treatment such as bleeding in sports and activities. So, the treatment is controversial. 3 On the other hand, APL positive in background autoimmune disorder such as SLE, aspirin is used although the strength of this treatment is controversial. Hydroxychloroquine an antimalaria medicine has an anti-inflammatory and prevent the clumping of platelet. It is used as supportive treatment in combination of special treatment in APS with difficult treatment of APS manifestations. Other immunosuppressive treatment especially should be considered in small vessel wall involvement with APL positive especially with damage of small vessel wall of the heart. In lupus patient with APS and bleeding usually has hypoprotrombinemia. Treatment with high dose corticosteroids, transfusion should be considered. In catastrophic APS (CAPS), high dose corticosteroids, IVIG with slower infusion, plasmapheresis and even rituximab should be considered. 4,5,6 References:





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### **Growing pain**

## ليلا شهبازنژاد $^1$ $^{ ext{C}}$

مرکز تحقیقات عفونی اطفال، پژوهشکده بیماری های واگیر، دانشگاه علوم پزشکی مازندران، ساری، ایران انجمن روماتولوژی کودکان رepdiatric Infectious Diseases Research Center, Communicable Diseases Institute,

Mazandaran University of Medical Sciences, Sari, Iran Pediatric Rheumatology society of Iran, Tehran,

Iran

Abstract: Growing pain (GP) is a common musculoskeletal problem in childhood; it estimated that nearly one third of children 4 to 6 years of old or 15% of school age children sometimes suffer from this pain. Most of the time the cases are between 3 to 12 years of old. GP is a non-inflammatory pain syndrome and is more prevalent than inflammatory limb pain in children. The pain most of the time occurs at night and in lower extremities in regions rather than joints, and most of the time is bilateral. There is not any back pain or point pain in the limbs. The pain may relieve by rubbing or drugs I.e. acetaminophen or other non-steroidal anti-inflammatory drugs. The pain is not a daily problem and may be on and off during a week or month. It may be worsened after an unusual increased physical activity. There is not any history of fever, weight loss or morning stiffness, on the other hand in the day after that night, the child is quiet healthy without any problem. Physical examination is completely unremarkable except for hypermobility joints in some cases. In conclusion growing pain is a diagnosis by exclusion and most of the time improved when the child get older. The physician has to pay attention to red flags and rule out the most serious differential diagnosis i.e. malignancies, infections or bone, joint or neuromuscular disease. Key words: Growing pain, children, nocturnal pain

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### **Hypermobility Spectrum Disorder**

Vadood Javadi, M.D 1 © P

**Abstract**: Generalized hypermobility is a starting point of a wide spectrum of generalized hypermobility. In this point, the child with hypermobility has not any complaint compatible with this problem. On the other hand, in the end point, the patient fulfills the definite Ehlers-Danlos syndrome (EDS) criteria. As definition, patients who do not complete the criteria called hypermobility spectrum disorder (HSD).(1) The most common complaint in HSD is musculoskeletal pain.(2) Some examples of other co-morbidities are fatigue, vaulting gait, flexible pesplanus, genu recarvatum, mitral valve prolapse, vesicoureteral reflux, primary focal hyperhydrosis, anxiety, dysautonomia, and phobia.(3) In this article the pathogenesis, epidemiology, classification, comorbidities and treatment points of hypermobility will be described. References: 1. Malfait. F, Francomano. C, Byers. P, Belmont. J, Berglund. B, et al. The 2017 international classification of the Ehlers-Danlos syndromes. Am J Med Genet Part C Semin Med Genet 175C: 8-26. 2. Parvaneh VJ, Shahvaladi H, Rahmani K, Yekta SJ, Gorji FA, et al. Correlation between benign joint hypermobility syndrome and primary focal hyperhidrosis in children: a novel concept. BMC Musculoskelet Disord. 2020 Apr 24;21(1):268. doi: 10.1186/s12891-020-03264-8. PMID: 32331513; PMCID: PMC7183110. 3. Javadi Parvaneh V, Modaress S, Zahed G, Rahmani K, Shiari R. Prevalence of generalized joint hypermobility in children with anxiety disorders. BMC Musculoskelet Disord. 2020 Jun 2;21(1):337. doi: 10.1186/s12891-020-03377-0. PMID: 32487116; PMCID: PMC7265217.





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## Secondary antiphopholipid syndrome in children (Rheumatologic causes)

نوید نمازی $^1$  ©  $^2$ 

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Abstract: Secondary antiphospholipid syndrome in children (Rheumatologic causes) Primary antiphospholipid syndrome occurs when no underlying disease presents, this type of APS maybe overestimated because some children later develop SLE. Thrombosis in children usually requires multiple risk factors to produce abnormal clothing, therefore clinical assessment of all children who have an APL-related thrombosis event should include a search for each disease with prothrombotic risk factor. Secondary APS results from underlying autoimmune diseases (most common, including rheumatologic diseases), Infection, Malignancies and Drugs. 50% to 70% of APS-cases in children are associated with an underlying autoimmune disease, specially SLE. Rheumatologic causes of secondary APS: I. SLE & SLE-like disorders (most common ) II. Rarely: Systemic vasculitis, JIA, Sjogren Syndrome, SSc, MCTD, IBD, JDM & ARF. Primary Vs Secondary APS Similarities (SLE-associated ): Positive ANA or anti-DsDNA, thrombocytopenia, nephritis, hemolytic-anemia and neuropsychiatric disorders . Differentiation between Primary Vs Secondary APS: Children with primary APS are Younger and have a higher frequency of arterial thrombosis. In contrast, children with APS associated with underlying autoimmune disease have a higher frequency of venous thrombotic events and hematological or skin manifestations. True vasculitis is rare in APS patients, then except in the setting of acute thrombosis, ESR and WBC are normal in APS patients, unlike SLE-associated APS cases. The prevalence of APL in children with SLE maybe significantly higher than adults, so the probability of primary APS become Secondary APS is higher in children, frequent evaluation of these children is necessary. Children with (JIA) have a high rate of APL but rarely develop APS (Non pathogenic APL ) . MIS-C , activates thrombosis cascade through different mechanisms, It remains unclear whether Apl are an epiphenomenon or are involved in the pathogenesis. Because of thrombophilic tendency in autoimmune rheumatologic diseases, it is recommended Apl be checked in all of this children. Thank you.





# The usage of a magnet-powered instrument in the extraction of metallic or magnetic aspirated foreign bodies: A case report

وزرخ دکتر مهدی صرافی دکتر غلامرضا ابراهیمی سراج دکتر محسن روزرخ  $^{1}$  ©

دانشگاه علوم پزشکی شهید بهشتی ۱

Abstract: Mehdi Sarafi, Mohsen Rouzrokh, Gholamreza Ebrahimi Saraj Pediatric Surgery Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran Introduction and importance The unintended passage of foreign bodies into the airway, is a common problem among children which can lead to death. Children may experience the sudden onset of symptoms like coughing, shortness of breath, cyanosis, wheezing, stridor, or choking. Prompt diagnosis and extraction of the entrapped object is the key of success in these cases. Rigid bronchoscopy traditionally has been the modality of choice for cases with AFB. Unfortunately, in addition to various extraction methods, in some cases the bronchoscopy may not be successful, and exploratory thoracotomy is inevitable. Case presentation We describe the story of a 7-year-old preschooler boy who was taken to the emergency department complaining the sudden onset of cough and dyspnea. He was conscious and stable at the first impression. His vital signs were within the normal range. Also, blood oxygen saturation was at 97 %. On physical examination, slightly reduced pulmonary sounds were detected on the right side. Initial investigations, showed an entrapped oval-shaped object in the right main bronchus. Several conventional bronchoscopic attempts including rigid broncoscopy were failed. Also, a posterolateral thoracotomy was performed on the right side which was inconclusive because of migration of the magnet during the procedure and inability to prolong the procedure due to nearly compromising hemodynamic and ventilatory status of the child. So, we decided to use a handmade magnet-powered instrument to extract the object endoscopically. Conclusion Using the magnet-powered grasping forceps may be beneficial after repetitive failed attempts in the extraction process of airway foreign bodies. Although there is no specific evidence-based guideline for choosing the best removal technique, we recommend using this technique in round-shaped high weight metallic objects as the

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## Which patients should refer to a Pediatric Rheumatologist?

Sahar Naderi Shiran <sup>1</sup> © P

Abstract: I.Inflammatory Joint Disease: Any patient with arthritis should be visited by a pediatric rheumatologist, especially if it is a chronic arthritis (6 weeks or longer). Patients with Uveitis also should refer to evaluate Juvenile Idiopathic Arthritis. 2.Inflammatory Connective Tissue Disease: When a patient has constitutional symptoms and multiorgan system involvement, we should evaluate systemic lupus erythematosus. Every child presenting with thrombosis, unexplained thrombocytopenia, hemolytic anemia, skin and neurological signs, and unexplained prolonged aPTT should refer to evaluate antiphospholipid syndrome. Heart block and skin rash in neonates also hepatic or hematologic abnormalities may be Neonatal Lupus Erythematosus, Child with skin rashes, photosensitivity, proximal weakness, nail fold erythema, calcium nodules and abnormal muscle enzymes should refer to evaluate Juvenile Dermatomyositis. Any Skin thickening, tightening, sclerodactyly needs evaluation of Scleroderma. Raynaud's phenomenon in patients with evidence of other organ involvement, Digital ulcers, positive ANA titer should refer. 3. Vasculitis: Unexplained fever, weight loss, fatigue, Skin and neurological symptoms, Arthralgia or arthritis, myalgia or myositis, serositis, hypertension, hematuria, renal failure, pulmonary symptoms, myocardial ischemia, arrhythmias, elevated inflammatory response, leukocytosis, anemia, thrombocytosis, hematuria and proteinuria should refer. 4. Autoinflammatory syndromes: Three or more episodes of unexplained fevers in a 6-month period, occurring at least 7 days apart and any periodic sign should refer to pediatric rheumatologist. 5.Infection Related Rheumatic Disorders: Peripheral Arthritis Predominantly lower limb, Plus Evidence of Preceding Infection within the 4 weeks prior should refer and evaluate reactive arthritis. In the case of Acute Rheumatic Fever, after a history of pharyngitis, when patient have Jone's criteria should refer. 6.Rheumatic Manifestations of Non-Rheumatic Disorders: In the case of osteoporosis and patients with musculoskeletal manifestations of systemic disease, generalized hypermobility, skeletal dysplasia, pain amplification syndromes, primary disorders of connective tissue should refer for further evaluation.





# A report of pediatric neuromuscular patients in Iran, a brief assessment of their diagnostic and treatment problems

MohammadKazem Bakhshandeh <sup>1</sup> © ®, Asghar Ghorbani

Abstract: Neuromuscular diseases are group of genetic muscle diseases that lead to wasting and loss of muscle. Hereditary neuromuscular diseases in children include a wide range of inherited diseases. In our country, due to the high rate of familial marriages (about 38-40%), hereditary types of pediatric neuromuscular diseases are common, which leads to an increase in the financial burden and health costs of families and the country's health system. Spinal Muscular Atrophy (SMA) and Duchenne Muscular Dystrophy (DMD) are most common hereditary neuromuscular diseases in children. Accurate and definitive diagnosis of hereditary neuromuscular diseases requires muscle biopsy and genetic analysis. Some gene mutation detection methods, such as Multiplex Ligation-Dependent Probe Amplification (MLPA), can be performed in Iran, but the investigation of point mutations in a series of pathogenic genes, such as Whole Exome Sequencing (WES) method, requires sending DNA samples to Germany, China, South Korea, etc. On the other hand, our country Iran is under the most severe economic sanctions of the enemies of humanity and the costs of sending genetic samples abroad are very expensive. The more unfortunate thing is that sometimes, despite these high costs and wasting the country's currency, the results of genetic analysis do not match the symptoms of the disease. Also many times the results are not confirmed without checking the samples of parents. Foreign pharmaceutical companies are doing extensive advertising in terms of new treatments, such as gene therapy about the healing properties of gene regulatory drugs and gene therapy. It is noteworthy that the costs of these gene therapy drugs are so high that they do not need to be reported. How is the efficacy of these expensive drugs? I believe that for pathological and genetic diagnosis and genetic treatment of pediatric neuromuscular patients, we need national decision along with self-confidence, relying on the





# 10-year follow-up report and management experience in a case of neonatal severe primary hyperparathyroidism



دانشیار ۱

**Abstract**: Background and aims: Neonatal severe primary hyperparathyroidism (NSHPT) is an extremely rare condition that appears in the first few weeks of a newborn's life in most cases. Additionally, it may adversely affect the neurological development of patients and even result in intellectual disability. Due to the importance of such complications, we present a 10-year follow-up and describe our experience in managing this case for the first time in Iran. Case presentation: We present the case of an 10-year-old boy who was diagnosed with NSHPT and underwent a total parathyroidectomy during his neonatal period. During the current follow-up, he exhibits mild intellectual disability and an epileptic disorder after ten years. Neonatal severe primary hyperparathyroidism (NSHPT) is a uncommon disease that in most cases becomes apparent during the first few weeks of a newborn's life. At 5 years old patient returned due to seizure, in work-up EEG demonstrated multiple epileptic discharges in a normal background and the patient was started on Carbamazepine. In spite of the use of this medication, seizures recurred twice when he was 7 and 9 years of age. In the present visit, weight and height of the patient were 30kg (50 percentile) and 132cm (10 percentile) respectively, which is normal .Patient's head circumference is 48cm (





## A common clinical and neuroimaging scenario with a different diagnosis!

Zahra Rezaei <sup>1</sup> © P

Abstract: Case Report: A 20 month infant who previously diagnosed to suffer from cerebral palsy, referred to pediatric neurology clinic for more evaluation. He was born to an elective caesarian section following an uneventful near term twin pregnancy. He and his brother experienced seizure at the first day of life. By considering twin near term pregnancy, hypoxia considered as the main etiology of neonatal seizure and neonates admitted in NICU for 2 weeks. The first brain sonography detected brain damages in favor of early onset hypoxia and the two brothers diagnosed as cerebral palsy. Generalized developmental delay followed by regression, poor controlled frequent seizures, consanguineous background as well as intensive brain cystic changes lead to suspicion about the underlying etiology. Pathogenic mutation in MOCS2 gene (Molybdenum cofactor deficiency of complementation group B) confirmed by genetic study and parent's segregation analysis. Conclusion: The lack of peri/pre natal insults, consanguineous background, and progressive disease course are some important clues irrespective of static nature of cerebral palsy. Key Words: Cerebral palsy, MOCS2, Molybdenum cofactor deficiency.





# An Introduction to the Griffith Mental Development Scales (GMDS-ER)

دكتر سيف اله حيدر آبادی  $^1$  ©  $^0$ 

دانشگاه علوم پزشکی تبریز ۱

**Abstract**: The core intention of studying normal child development and devising standardized tests of development is to better understand and plan appropriate therapeutic plan for children who are not developing normally. Originally, The GMDS published in 1954.. The GMDS – Extended Revised published in 2006. They are used for evaluating the developmental skills of a child in several developmental domains. It measures the rate of development of infants and children from birth to eight years and has 2 different age groups packages, including 0-2 and 2-8 years old packages. The GMDS assess a child's strengths and weaknesses in 5 developmental skills (0-2 package) and 6 developmental skills(2-8 years) and determines if a child should receive early intervention. Skills assessed by GMDS are as follows: A. LOCOMOTOR SCALE: Gross motor skills, including the ability to balance and to coordinate and control movements. B. PERSONAL-SOCIAL SCALE: Self help skills in eating, dressing, personal hygiene and domestic activities; social responsiveness to surroundings, socialization, play. C. HEARING AND LANGUAGE SCALE: Responsive and active listening, speech, receptive and expressive language, comprehension and verbal concepts, D. EYE AND HAND CO-ORDINATION SCALE: Fine motor ability and coordination, reaching, grasping, manipulation of small objects and pencil use. E. PERFORMANCE SCALE: Active application of fine motor skills to purposeful activity with objects, non- verbal problem solving F. PRACTICAL REASONING SCALE: Starts in Year III. Development of number and size concepts, time, understand basic maths concepts, moral reasoning, auditory and visual short term memory. The scores obtained from GMDS are row scores, age equivalents, Sub-Quotients and General Quotient, Percentiles





### ASQ-7, a Valid Children's Developmental Screening Tool

### Bahar Allahverdi MD Pediatric Gastroenterologist <sup>1</sup> © P

Abstract: Screening young children is an efficient way for professionals to check a child's development, help parents celebrate their child's milestones and know what to look for next, and determine whether follow-up steps are needed. It's also an essential first step toward identifying children with delays or disorders in the critical early years, before they start school. Evidence shows that the earlier development is assessed the greater the chance a child has to reach his or her potential. The Ages & Stages Questionnaire 3rd edition, (ASQ-3) is a developmental screening tool that pinpoints developmental progress in children between the ages of one month to 5 ½ years. Its success lies in its parent-centric approach and inherent ease of use, a combination that has made it the most widely used developmental screener across the globe. ASQ-3 relies on parents as experts, is easy-to-use, highly valid, reliable, and accurate and creates the snapshot needed to catch delays and celebrate milestones. It highlights a child's strengths as well as concerns. The questionnaires are available in multiple languages. In IRAN, ASQ-3 has been validated by experts and is available in Persian. ASQ-3 captures parents' indepth knowledge, takes just 10 to 15 minutes for parents to complete and 2 to 3 minutes for professionals to score. It can be completed at home, in a waiting room, during a home visit, or as part of an in-person or phone interview. In terms of being family-friendly, ASQ-3 is costeffective, free of charge in urban and rural public health care centers, teaches parents about child development and their own child's skills and highlights results that fall in a "monitoring zone," to make it easier to keep track of children at risk. Surprisingly, ASQ-3 has lot of fun and is engaging for kids.





### Autistic spectrum disorder

## Hadi Montazerlotfelahi <sup>1</sup> © P, Keyhan Mostafavi <sup>2</sup>

**Abstract**: ASD is not a disease, it is a developmental behaviorally syndrome that impacts social skills and verbal-nonverbal communication with narrow, rigid, rigid, repetitive behaviors and interests. ASD affect the immature, developing brain. Epilepsy, macrocephaly, speech disorders, sleep problems are exist with ASD. Network dysfunction and neurotransmitter disorders is pathogenesis of ASD. Male to female ratio is 4.5:1. Mean age diagnosis ranged from 3.5-5 yrs. Early diagnosis and intervention is critical, occupational therapy, speech therapy and medical treatment is helpful. Treatment of comorbiditeis( ADHD, angsiety, tic, OCD...) is important.





### Cerebral Palsy overview and its classifications

### Mahmoud Reza Ashrafi <sup>1</sup> © P, Ali Reza Tavasoli <sup>1</sup>

**Abstract**: Developmental disabilities are a group of related early onset, nonprogressive and chronic neurologic or behavioral disabilities occurring in childhood and estimated to affect 5-10 % of children. Cerebral Palsy (CP) is a developmental disability in the field of motor skills. The first description of cerebral palsy as a clinical entity is attributed to William John Little, an eminent British orthopedic surgeon. CP is now familiar to most health and social service professionals, as well as many members of the general public, as a physically disabling condition. The term 'cerebral palsy' was defined as "a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain" (2007 consensus definition). Cerebral palsy is the most common cause of physical disability in childhood. The worldwide prevalence of cerebral palsy has remained stable at 2–3 per 1000 livebirths for more than four decades, despite substantial improvements in obstetric and neonatal care. Cerebral palsy (CP) is a non-progressive neurodevelopmental disorder characterized by motor impairments, often accompanied by co-morbidities such as intellectual disability, epilepsy, visual and hearing impairment and speech and language deficits. Cerebral palsy can be classified on the basis of four major components: type and severity of the motor abnormalities, anatomical distribution, associated impairments, and timing of the presumed causal event (prenatal, perinatal, or postnatal). Although there is a clear role for hypoxic-ischemic injury in some cases of CP, estimates suggest that acute intrapartum hypoxia-ischemia accounts for fewer than 10% of cases. Some investigators suggest that 'unknown pathophysiologic processes' must be at work to account for a significant proportion of CP. Much of this unknown pathophysiology may be owing to genetic or epi-genetic factors. Indeed, current estimates indicate that as many as 30% of CP cases





### Clinical approach to the dysmorphic child

### Dr Maryam Sotoudeh anvari <sup>1</sup> © ®

Abstract: The child affected by a Dysmorphology represents a care challenge for the pediatrician. He is in fact the heart of the multidisciplinary team that has to manage the patient, trying to control the complications of his/her syndrome and promoting the correct psychophysical development. Around 4000 malformation syndromes have now been delineated. The approach to making a diagnosis of a dysmorphic child is fundamentally similar to making a diagnosis of a child with any systemic illness and relies on a detailed history, a meticulous clinical examination for detailing the major and minor anomalies, recording the growth, examination of previous records and photographs are complemented by cytogenetics and molecular genetic techniques in achieving a diagnosis. A genetic etiology should be suspected if the child /infant has any of the following: a. Congenital anomalies: one or more major anomaly or more than two minor anomalies b. Poor growth: symmetric intrauterine growth restriction or postnatal growth failure c. Developmental delay or developmental regression d. Craniofacial dysmorphism e. Ambiguous genitalia Familiarity with dysmorphology databases and cross referencing the anomalies especially the rarer ones helps in narrowing the differential diagnosis.





# Clinical features and differential diagnosis of feeding and eating disorders in children and adolescents

الهام سالاری $^1$   $^{f C}$   $^{f P}$ 

بیمارستان روزبه <sup>۱</sup>

Abstract: Because eating disorders have recently become more prevalent among younger patients, it is incumbent upon pediatric health care providers to recognize the signs and symptoms and to make prompt diagnoses or refer to specialists as necessary. The medical findings associated with eating disorders such as anorexia nervosa in children and adolescents are usually similar to those in adults, with some exceptions. First, children and adolescents may become medically compromised much more rapidly than adults because of reduced nutritional reserves and increased metabolic demands for growth and development. Thus, significant medical complications can occur with a smaller relative amount of weight change or in the context of rapid weight loss. Second, certain complications such as growth retardation, interruption of puberty, and interference with peak bone mass acquisition and brain development have a greater impact in children and adolescents and are potentially irreversible. As a result, medical professionals should be able to identify eating problems in children and adolescents in its early stages. However, the diagnosis of an eating disorder in this age group can be particularly challenging because these patients frequently fail to endorse cognitions typically associated with eating disorders (e.g., feeling fat, fearing weight gain, concern about body shape or weight) but may instead present with vague physical complaints such as nausea, difficulty swallowing, or abdominal pain after eating. In this lecture we will provide instructions for conducting a careful history and physical examination, evaluating physical findings associated with anorexia nervosa and bulimia nervosa in children and adolescents, confirming an eating disorder diagnosis by excluding other causes of weight loss or vomiting, and indications for hospitalization in a child or adolescent with an eating disorder.





## Clinico-Neurophysiological Features Of ESES/CSWS

## Alireza Rezaei <sup>1</sup> © P

Abstract: Electrical status epilepticus during slow sleep (ESES) or continuous spikes and waves during slow sleep (CSWS) is a condition characterized by presence of epileptiform activity in the electroencephalogram (EEG) during sleep. The aim of this study is to analyze neurophysiological and clinical characteristics of patients with CSWS, in order to describe a clear method for diagnosis of ESES by a routine EEG or Video-EEG-Monitoring. ESES is an EEG pattern that appears during childhood, and is often associated with cognitive impairment. It can appear in the course of epileptic syndromes such as Landau-Kleffner syndrome (characterized by acquired aphasia presenting with or without epileptic seizures), atypical benign partial epilepsy of childhood (infrequent forms of benign epilepsy manifesting with atonic seizures, nocturnal sylvian seizures and sometimes absences, behavioral problems and cognitive deterioration) and benign epilepsy of childhood with centrotemporal spikes (focal epilepsy with benign sylvian seizures at night, in the context of which CSWS may develop in exceptional cases). There are not enough series studies but a lots of case series were presented. Definitions of ESES is vary and the diagnostic process is complicated. Some studies focus on quantitative method, however, qualitative methods were presented in others. For clinical purposes, a nap EEG after sleep deprivation without sleep medication combined with an EEG during the awake stage probably will suffice. If there is a high suspicion of CSWS, one could expand the registration to a longer period, preferentially a 24-h EEG. The criterion of at least 50% epileptiform activity during non-REM and/or REM sleep seems most adequate, especially if the clinical picture fits a CSWS/ESES-related syndrome. KEY WORDS: EEG, Epilepsy, Landau-Kleffner syndrome, SWI, Cognitive decline.





### **Cognitive Development**

## دكتر محمد وفائي شاهي $^1$ $^{\odot}$

مرکز تحقیقات رشد و تکامل کودکان، انسیتو غدد درون ریز و متابولیسم، دانشگاه علوم پزشکی ایران ۱

**Abstract**: Cognitive development is a field of study in neuroscience and psychology focusing on a child's development in terms of information processing, conceptual resources, perceptual skill and language learning. Cognitive development is defined as the emergence of the ability to consciously cognize, understand, and articulate their understanding. Cognitive development is how a person perceives, thinks, and gains understanding of their world through the relations of genetic and learning factors. There are four stages to cognitive information development. They are, reasoning, intelligence, language, and memory. Jean Piaget was a major force establishing this field, forming his theory of cognitive development. Piaget proposed four stages of cognitive development: the sensorimotor, preoperational, concrete operational, and formal operational period. Many of Piaget's theoretical claims have since fallen out of favor. Within the constructivist approach, an exciting new set of studies and theoretical ideas confirms something that parents and preschool teachers and others have long thought intuitively: Very young children's play, both their exploratory play and their imaginative and pretend play, contributes greatly to their early learning. Nativism, empiricism, and constructivism all focus on the process of learning from evidence. Two other approaches describe other factors that contribute to cognitive development. Information-processing approaches stress the development of general abilities to process and organize information, such as memory or attention. Indeed, children do develop such abilities in the first few years of life and those developments contribute to the development of their knowledge. Within the 1st mo infants prefer to look at human faces and listen to human voices, and rapidly prefer the face, voice, and even smell of their caregivers. Within the 1st vr babies develop an even richer understanding of others. In their second year, children also start to understand that their own perceptions, attention, and emotion may be shared by others.

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# Common reactions of children and adolescents to illness and hospitalization

غزال زاهد  $^1$  ©  $^{ ext{P}}$ 

**Abstract**: Having a serious, chronic, and debilitating illness, as well as long-term and frequent hospitalizations, increases the likelihood of subsequent psychological problems. Younger children, especially between 6 months and 4 years, are more vulnerable, mainly due to disturbance in daily routine and usual activities, and lack of communication with peers. Unfamiliar environment and uncertainty about what are going to happen (a set of unknowns, a strange environment and treatment personnel) causes anxiety in child. Although most children have experienced hospitalization in the past, familiarity with hospital routines and/or previous treatments does not necessarily eliminate or reduce a child's reaction to hospitalization, and most children are nevertheless afraid of the personnel and treatments they will have to endure. Common stressors are: separation from parent and fear of abandonment, unfamiliar environment, loss of autonomy and mobility, change in routine, limited caregiver involvement, and Over-stimulation/ under-stimulation. Some children have difficulty sleeping due to poor ventilation, bright lights at night, and noise caused by ringing phones, nurses talking, excessive heat in the environment, and crying of other children. An increased level of preoperative anxiety increases the likelihood of negative psychological outcomes during the recovery period, such as: pervasive anxiety, separation anxiety, sleep problems, nightmares, irritability, and aggressiveness. Reactions vary by age and developmental level. Despite these issues, most children show adaptation when it comes to "getting used" to the unfavorable aspects of the hospital environment.





# Cultural adaptation, validation and standardization of the developmental screening tools ASQ: SE-2 in Iranian children

# Roshanak Vameghi <sup>1</sup>, Hamid Reza Lornejad <sup>2</sup> ©, Naria Abolghasemi <sup>2</sup> ®

Abstract: Summary: The present study aimed at culturally adapting, validating and standardizing the ASQ:SE-2 by implementing a nation-wide cross-sectional methodological study, in order to provide a valid and reliable tool for determining the developmental status of Iranian children. Material & Methods: ASQ-SE consists of 9 questionnaires for 2, 6, 12, 18, 24, 30, 36, 48 and 60 months age groups. Each questionnaire has 19 to 33 questions (with different number of questions for each age group). Also at the end of each questionnaire there are open-ended questions allowing parents to express concerns regarding their child. After translation and back-translation of tools, they were validated formally and conceptually their cross-cultural adaptation were assessed by 51 specialists and experts related to the field of pediatrics and child development. Then, in order to implement a cross-sectional study, the country was divided into 14 areas and a multi-stage cluster sampling method was used to recruit the sample. In each area, 10 urban and 10 rural clusters (with 21 girls and 21 boys in each cluster: 1 girl and 1 boy for each age group in each cluster) were randomly selected. The reliability, cutoff points for each age group were determined. All statistical analyses were performed. Results: In the cross-sectional study, a total number of 11740 children aged 1-66 months were studied. In terms of the ASQ:SE-2, based on the results derived from performing these questionnaires on all research subjects in all age groups, the overall prevalence of socialemotional developmental delays (scores higher than the cut-off) was 22.8% and 7% of all children scored in the monitoring zone. The results of this study showed that the Iranian version of ASQ:SE-2 is valid and reliable; moreover, the cut-off points designated for it can be implemented in the Iranian children community to assess their developmental status.





### **Development and sex education**



بیمارستان روزبه ۱

**Abstract**: Developing a healthy sexuality is a key developmental milestone for all children and adolescents. Sexuality education is more than the instruction of children and adolescents on anatomy and the physiology of biological sex and reproduction. It covers healthy sexual development, gender identity, interpersonal relationships, affection, sexual development, intimacy, consent, and body image for all adolescents, including adolescents with disabilities, chronic health conditions, and other special needs. Healthy sexuality includes the capacity to promote and preserve significant interpersonal relationships; value one's body and personal health; interact with both sexes in respectful and appropriate ways; and express affection, love, and intimacy in ways consistent with one's own values, sexual preferences, and abilities. Healthy sexuality is influenced by ethnic, racial, cultural, personal, religious, and moral concerns. Information about sexuality can be taught and shared in schools, communities, homes, and medical offices using evidence-based interventions. As a result, the pediatrician should encourage early parental discussion with children at home about sexuality. Pediatricians should model ways to initiate talks about sexuality with children at pertinent opportunities and how to answer children's questions fully and accurately, based on the child's age. Moreover, parents should be taught how to introduce puberty to their pre-teens and handle the related changes. Besides, the prevention of child sexual abuse is widely based on child-centered education, teaching children to identify, avoid and report sexual abuse. Parents should be informed how to be the protectors of their children through supervision, monitoring and involvement as well as the promotion of the self-efficacy, wellbeing, and self-esteem of their children. We will discuss these issues in more details in this lecture.





### **Disclosing an Adverse Event or Medical Error**

عفت حسينعلى بيگى  $^1$  نيلوفر قنبرى  $^{ ext{@}}$  فاطمه خانى نوذرى  $^2$ 

**Abstract**: When an unanticipated adverse event, such as a medical error, occurs, disclosure of the event, to the patient and family is required ethically: as part of a professional obligation to the patient and family; to comply with regulatory requirements; and, depending on the state in which one practices, to be in compliance with statutory requirements. The inclusion of an apology with the disclosure of an adverse event in the healthcare setting is an area of active debate, which will be discussed further in this chapter. Internal reporting, such as through a facility's adverse event reporting system, is not addressed in this paper. Definitions: Medical Error: An act of commission (doing something wrong) or omission (failing to do the right thing) that leads to an undesirable outcome or significant potential for such an outcome. Near Miss: An event or situation that did not produce patient injury but only because of chance. Adverse Event: Any unanticipated, negative event that occurs during the care of a patient regardless of circumstance. Some are preventable, others are not readily avoidable. Disclosure: Telling the patient and family about an adverse event is a disclosure. Disclosure typically includes a statement of recognition that an adverse event has occurred and an explanation of what is known about how the adverse event happened. Apology: An apology is an expression of regret and compassion that an adverse event has occurred. What are the barriers to disclosing adverse events? The American Society for Healthcare Risk Management (ASHRM) (2003) characterizes barriers to disclosing adverse events to patients and families as either being rooted in personal beliefs or fears or based on perceptions of the legal process that may ensue after the disclosure is made.





### **Disclosing and Adverse Event or Medical Error**

عفت حسينعلى بيگى  $^1$  فاطمه خانى نوذرى  $^2$  نيلوفر قنبرى  $^2$ 

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**Abstract**: What do patients and families do? For both moderate and severe medical errors. patients were significantly more likely to consider bringing a suit against the physician if the physician failed to disclose the error. Specifically, 12% of patients would bring a lawsuit against the physician if they were informed, while 20% would sue if the physician did not disclose and the patient discovered the error by some other manner. Guiding principles for disclosing adverse events require that: • All caregivers have the patient's best interest in mind • The physician is ultimately responsible for treatment decisions • The physician and the organization are responsible for providing quality patient care • Performance improvement and patient safety are continuous tasks Nationally Recognized Models to Address Disclosure 1)Extreme Honesty 2)Open Disclosure with Offer: Three principles guided this new systematic approach to adverse events: compensate patients quickly and fairly when unreasonable medical care causes harm; if the care is deemed reasonable, support caregivers and the organization vigorously; reduce patient injuries by learning through patients' experiences and also reduce claims (by way of improved care). 3) Seven Pillars: The pillars are: 1) Reporting 2) Investigation 3) Communication 4) Apology with remediation (includes waiving of hospital and physician fees) 5) Process and performance improvement 6) Data tracking and analysis 7) Education around the entire process 4) A Practical Guide to Disclosure: models for adverse event disclosures can be summarized as: • One person alone (aka, the Lone Ranger approach) • Team oriented – Small group setting – Large group setting 5)Conducting the Disclosure 6)Closing the Disclosure. Conclusion Disclosure of a medical error to the patient and family affected by the error is required ethically, as part of a professional obligation to the patient.

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poster

# Factor structure and psychometric properties of a Persian translation of the Epworth Sleepiness Scale for Children and Adolescents

ويدا ايماني  $^1$  ©  $^2$ 

**Abstract**: Given the high prevalence of excessive daytime disorder (EDS) among children and adolescents, daytime sleepiness should be effectively measured for them to design appropriate intervention program. However, the commonly used instrument Epworth Sleepiness Scale for Children and Adolescents (ESS-CHAD) has little information in its psychometric properties. This study aimed to apply 2 different test theories to examine the psychometric properties of the Persian ESS-CHAD among a large sample of Iranian adolescents and children. Methods: In this methodological study, participants from 8 high schools (n= 1371; 700 males), in Qazvin, Iran, completed the ESS-CHAD, a background information sheet, and Insomnia Severity Index (ISI). The ESS-CHAD was translated by using a forward-backward translation method. Two weeks later, the participants completed the ESS-CHAD again. Internal ...





# Genetic Diagnosis of spinal Muscular Dystrophy

Morteza Heidari <sup>1</sup> © P

**Abstract**: Genetic Diagnosis of spinal Muscular Dystrophy Spinal muscular atrophy (SMA) is an inherited neuromuscular disorder that causes degeneration of the anterior horn cells in the spinal cord, which causes severe progressive hypotonia and muscular weakness. With a carrier frequency of 1 in 40–50 and an estimated incidence of 1 in 10,000 live births, SMA is the second most common autosomal recessive disorder. Affected individuals with SMA have a homozygous loss of function of the survival motor neuron gene SMN1 on 5q13. The most common mutation causing SMA is a homozygous deletion of the SMN1 exon 7. Because SMN2 produces a reduced number of full-length transcripts, the number of SMN2 copies can modify the clinical phenotype and as such, becomes an essential predictive factor. Population-based SMA carrier screening allows the carriers to make informed reproductive choices or prepare for immediate treatment for an affected child. Newborn screening for SMA has been shown to be successful in allowing infants to be treated before the loss of motor neurons and has resulted in improved clinical outcomes. Keywords: spinal muscular atrophy, carrier screening, newborn screening





## Genetic diseases causing cerebral palsy

# Alireza Biglari, MD PhD <sup>1</sup> © P

Abstract: Genetic diseases causing cerebral palsy Alireza Biglari, MD PhD Children's Medical Center, Tehran University of Medical Sciences, Tehran, IRAN Cerebral palsy is a heterogeneous condition with diverse clinical manifestations and different brain imaging patterns, underlying causes, and more recently heterogeneous genetic etiologies. Previous studies have shown that hereditary factors can predispose an individual to cerebral palsy through gene-to-gene interactions or complex interactions with multiple environmental influences. Some of the genetic roles in causing abnormal conditions during pregnancy include placental abruption, fetal growth restriction, chorioamnionitis, premature birth, and preeclampsia. In recent years, modern diagnostic tools, such as genome sequencing, have enabled more detailed and comprehensive investigations and provided the basis for proving the direct role of several known gene mutations as an underlying cause in some cerebral palsy patients with the potential to change the approach to their care. These mutations often interfere with early brain development or predispose individuals to certain environmental risk factors. Based on the results of published studies, genetic evaluation including genome sequencing should be considered for any child with cerebral palsy without known risk factors, or whose condition worsens over time. Also, if there is a birth defect in the child or other family members, genetic testing in children with known risk factors for cerebral palsy can be recommended. The identification of genetic causes of cerebral palsy can provide the basis for the development of interventions based on pathology.





#### imaging in cerebral palsy

Neda Pak <sup>1</sup> © P

Abstract: Imaging in Cerebral palsy Cerebral Palsy (CP) is a clinical diagnosis, based on neurological symptoms and a motor disorder. Although neuroimaging is not part of definition of CP, neuroimaging is abnormal in more than 80% of cases with CP, determining the pathogenic pattern responsible for the CP and MRI is the first diagnostic modality. Brain abnormalities in CP arise at different times during brain development.in patients with preterm birth, white-matter damage, including PVL is the most common finding presenting as white mater loss and gliosis, followed by basal ganglia lesions, cortical/subcortical lesions, focal infarcts Normal MRI findings is also present and normal MRI doesn't exclude the CP diagnosis. This type of damage mostly occurs before 34 weeks of gestation but could also be seen in term infants. Basal ganglia and thalamic damage is mainly associated with dystonic CP, and usually happens in more sever hypoxia. mild hypoxia in term patients usually manifest as abnormality in perirolandic regions. MRI can help clinicians to answer the question whether the brain damage of the newborn is responsible for its clinical condition, MRI can also contribute to determining the prognosis of the infant's future development. advanced imaging such as functional MRI and DTI could be helpful in evaluation of prognosis of CP patients.





## Juvenile dermatomyositis

سعید انوری $^1$  ©  $^2$ 

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Abstract: Dr. Saeed Anvari Pediatric Neurologist Social Security Organisation Juvenile dermatomyositis or JDM JDM is an inflammatory disease of the muscle and skin and blood vessels. Patient with JDM have varying symptoms ranging from mild muscle weakness like difficulty getting out of a chair or difficulty turning over in bed to severe symptoms including profound weakness or difficulty swallowing. Fatigue rather than the specific complaints of muscle weakness is a common presenting complaint in JDM. Dermatomyositis is a rare autoimmune inflammatory myositis of unknown etiology effecting both children and adults. It involves strained muscles and skin. Unlike the adult form it does not have an increased risk of malignancy. JDM present at any age including infancy although most cases occur between age five and fortteene years. JDM common is approximately two to four children per one million children. Our case was a three year old and three month old boy who had been weak and lethargic for about a year before his visit. He developed thrombocytopenia two months after the onset of weakness and with the diagnosis idiopathic thrombocytopenia purpura. He had received IVIG and he received prednisolone for about one month for an unknown reason. At the time of our first visit the patient was weak and a little lethargic and restless and during the examination he was difficult to get up from the floor that is gower sign was positive. The environmental connection and cognition was appropriate with age. Our case did not have heliotrop rash and gottron paperless and abdominal pain or dysphagia or fever and calcinosis. EMG and NCV was nonirritble myogenic changes. And finally a muscle biopsy was performed for the patient that had inflammatory cells around fascicle and between fibers and dermatomyositis was confirmed. After prescribing methylprednisolone we started oral prednisolone with azathioprine.





# LAMA2-related muscular dystrophy mimicking spinal muscular atrophy

Azita Tavasoli <sup>1</sup> © P

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Abstract: Laminin-alpha 2-related dystrophy (LAMA-MD) is a muscular dystrophy due to reduced Laminin subunit α2, an extracellular matrix protein that links with dystrophin on the inner side of the muscle membrane. It has a heterogenous disease spectrum ranging from a severe, early onset congenital muscular dystrophy (complete Laminin subunit α2 deficiency, also called merosin-deficient congenital muscular dystrophy type 1A) to a mild, childhood- or adult-onset limb-girdle type muscular dystrophy (partial Laminin subunit α2 deficiency). Moreover, patients may develop epileptic seizures and may show characteristic diffuse brain white matter lesions on magnetic resonance imaging. Laminin alpha 2 deficiency can be secondary to defects in other proteins involved in the dystroglycan complex or pathway. Early onset disease is presented at birth or within the first 6 months of life with profound hypotonia. poor spontaneous movements and large joints contracture, feeding difficulties and respiratory failure. Intellectual function usually is normal. In later onset form, cardiac involvement and demyelinating sensorimotor neuropathy may be developed. In the first year of life, serum ceratine kinase level may be more than fourfold normal. In adults, CK is only mildly elevated. Structural brain abnormalities secondary to migration defect could be seen on MRI included cortical dysplasia, lissencephaly and polymicrogyria. Immunohistochemistry findings of muscle or skin biopsy and muscle MRI are also helpful. The diagnosis is confirmed by detecting recessive pathogenic variations in LAMA2 gene. The disease must be distinguished from other congenital muscular dystrophies, congenital myopathies, congenital myasthenic syndromes, and spinal muscular atrophy. There is not any treatment option for the disease. optimal symptomatic therapy in combination with rehabilitation is essential. promising new therapies are currently being developed including preclinical studies on the use of linker proteins, exogenous administration of Laminin-111, upregulation of LAMA1, genome editing technology and the use of antioxidant molecules.





# Maternal care system during the children's hospital admission

بهار اله وردى  $^{1}$   $^{0}$ 

**Abstract**: When a child is hospitalized, the family becomes especially vulnerable and scared. During this period, they need special support. Despite the fact that the healthcare providers sympathize with the mothers, they still cannot fully understand them, having not been in their place. Many aspects of the parents' life will change during hospital stay, including their natural needs and social and economic issues, which can cause stress and anxiety for the parents. Feelings of stress and anxiety are often associated with the lack of information on diseases and medical procedures. The pain is caused by the imposed treatments, unfamiliarity with the hospital rules and regulations, unfriendly staff and being afraid of asking questions. In addition, a higher level of family stress can reduce the ability of the mother to cope with problems during her child's hospital stay. Despite the importance, in most hospitals there are no action plans or training programs to reduce stress for the parents. Hospital staff and parents have different perceptions of stressors in the child's admission to a hospital. Because of staff familiarity with the hospital environment, they do not assume that the hospital environment and setting can be a stress causing factor for the mothers of hospitalized children. In other words, efforts that the physicians and nurses make to reduce stress for parents may not be sufficient and on the contrary can increase their stress levels. It is important to identify the stressors in the parents of hospitalized children and their impact on the treatment process and also to find out how these factors may be affected by different cultural background, ethnicity and the region. Special attention should be given to identify the stressors in nursing care, planning and parent education, moving stressors and preparing the best treatment facilities in the same direction.





## **Neuroscience in pediatrics**

دکتر منصور بهرامی  $^1$  ©  $^0$ 

دانشگاه علوم پزشکی شهید بهشتی ۱

Abstract: Developmental neuroscience has become increasingly relevant to clinical child neurology so that it is important to include selected areas of this knowledge into paediatric clinical practice. It is now very common for some pediateric conferences committees to have some presentations that deal with the pathogenesis of neurological disorders and discuss about the molecular aspects of neurological diseases. Accordingly it is important for paediatricians to be involved in such presentations and group discussions of the relevant neurobiology literature and when possible some exposure to hands on research. In this presentation I will be discussing the importance of neuroscience in development paediatrics and its vital role to understand the etiology of some disease such as Autism and ADHD. Dr. Mansour Bahrami MD





#### **Parenteral Communications**

Iran Malekzadeh <sup>1</sup> © P

Abstract: Family is our first perception of social communication and almost always this group membership lasts our whole life, every day, and in every emotional experience. During the first days, kids can only communicate through crying; which can mean a variety of needs, such as thirst, hunger, or pain. Parents' responses must be quick and profound to create a sense of love and safety in the child. In the first months, children can smile and follow with their eyes, which is the initiation of social active responses. Interactions improve step by step and parents' reactions, as reward or punishment, will encourage or discourage the child's activities. After the first year, children can speak and parents can guide and respond to their emotions, more easily. Triangular communication starts at 3 years old when the children understand the meaning of emotional interactions and the role of the parents. Parents should create children's structure of feelings, security or danger, being loved or unwanted, freedom or restriction.





# Right transvers sinus Thrombosis after COVID-19 in 14 years old boy

ساسان ساکت  $^1$   $^{\odot}$   $^{\odot}$ 

**Abstract**: Abstract Background: Cerebral sinovenous thrombosis (CSVT) is a rare but serious disorder affecting children from the newborn period till 18 years old. The annual incidence of CSVT is approximately 0.6 per 100,000. Coagulation disorders also increase the risk of CSVT, which can result in ischemia and hemorrhage as the cerebral venous pressure rises. CSVT can also be caused by infections or dehydration. Case presentation: The child, a 14-year-old male, presented with a history of seizure, lethargy, weakness, and right sided hemiparesis with the final diagnosis of CSVT due to COVID-19 that is a rare condition among young boys. He finally discharges with anticoagulant and anti-seizure drug and advise to perform neurorehabilitation. We follow-up him for more than 2 months. Conclusion: During the COVID-19 pandemic, in patients presenting with neurological manifestations of CVST, it is advisable to look for a current or recent infection of COVID, regardless of the presence of respiratory symptoms. Keywords: Pediatric, Stroke, Cerebral sinovenous Thrombosis, COVID-19





## Screening tests in healthy children

#### Golnaz Ghazizadeh essalmi <sup>1</sup> © P

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**Abstract**: For healthy newborns on the day 3 to 5 after the birth, the blood spot specimen is obtained by puncture of newborn's heel, in order to detect hypothyroidism, phenylketonuria and G6pd deficiency Oral health risk assessment begins at six-month-old and referral to a dental home should be at last at one year old. The AAP suggests universal laboratory screening for Iron deficiency anemia at approximately one year of age and repeated screens for children with risk factors. Hearing loss screening in Iran is recommended early after birth before discharge from the hospital and if not available by the end of first month of life. Diagnosis of any auditory disorders before 3 months and intervention before 6 months of age is highly suggested. Selective hearing loss screening is recommended in 12, 15, 18, 24, 30 months of age in children with risk factors such as prematurity, congenital infection, hyperbilirubinemia, meningitis and expose to ototoxic drugs. Periodic vision screening is aimed to detect amblyopia, strabismus, and other vision problems. In children younger than 2 to 3 years of age, visual behavior and for those older than 3 years, visual acuity is assessed. We screen children without risk factors or conditions associated with hypertension by measuring blood pressure annually at health supervision visits, beginning at age three years. For children < 3 years with risk factors such as recurrent urinary tract infection, proteinuria, renal disease or urologic malformation, family history of congenital renal disease, and solid organ or hematopoietic cell transplant, we measure blood pressure at all health supervision visits. Screening for lead poisoning should be considered in regions with high prevalence, in children 9 to 12 months and 24 months old. For children without findings associated with Autistic spectrum disorder (ASD), we suggest ASD-specific screening at 18 and 24 months of age.





# Severe recurrent Acute Necrotizing Encephalopathy (ANE) due to covid-19

# Simin Khayatzadeh Kakhki <sup>1</sup> © P

Abstract: Background: Acute necrotizing encephalopathy (ANE) is a rare but rapidly progressing encephalopathy following a febrile illness, commonly a viral infection. It is characterized by the features of acute encephalopathy such as seizure, alteration of consciousness, and symmetric involvement of the bilateral thalamus on neuroimaging tests. Although most ANE cases have occurred sporadically, familial or recurrent ANE has been reported in Caucasian patients, with genetic susceptibility to ANE noted in some patients due to a RANBP2 mutation. We report the cases of two Korean siblings with typical ANE and RANBP2 mutation. Case report: A 5.5-year-old girl of nonrelative parents presented with deterioration and loss of consciousness after three days of fever and positive COVID-19 PCR test. Brain magnetic resonance imaging scan (MRI) showed multiple abnormal signal foci at periaqueductal region, bilateral thalami, external capsules associated with diffuse abnormal signal at midbrain, pons, and medulla. After she received intravenous immunoglobulin and high-dose corticosteroid pulse; she has got better, but had not return to normal background. She had a previous history of similar event when she was 6-month-old. In the end, she was diagnosed with familial ANE after identifying a mutation in RANBP2 (c.1754C>T: p. Thr585Met). Conclusions: This is the interesting case of recurrent familial ANE identifying a RANBP2 mutation. To recurrent nature of familial ANE and progressive deterioration, genetic test of RANBP2 mutation should be considered for early diagnosis in any cases of acute necrotizing encephalopathy of childhood. Keywords: recurrent encephalopathy, Acute necrotizing encephalopathy; Familial; RANBP2.





# Spinal muscular atrophies Classification

# Mahmoud Reza Ashrafi <sup>1</sup> © P, Morteza Heidari <sup>1</sup>, Ali Reza Tavasoli <sup>1</sup>

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Abstract: Spinal muscular atrophies (SMAs) are genetic disorders that are clinically characterized by progressive muscle weakness, hypotonia and atrophy, associated with degeneration of the alpha motor neurons from anterior horn cells in the spinal cord and, in the most severely affected patients, lower bulbar motor neurons. Classic proximal SMA is the most common form of SMA and the leading cause of infant mortality; it seems to be found in practically all populations but is diagnosed more frequently in infants and children, rather than adults. Although SMA is considered a rare disease and the global incidence of live births is estimated to be 1/6000 to 1/10,000, SMA is still the second most common autosomal recessive genetic disease and the most common monogenic disorder that causes early infant death. Over 95% of people with SMA harboring a homozygous deletion of the Survival of Motor Neurons1(SMN1) gene located on chromosome 5q13. The survival motor neuron (SMN) gene comprises nine exons and has been shown to be the primary SMA-determining gene. Humans have 2 nearly identical inverted SMN genes on chromosome 5q13 .SMN1, the telomeric copy of the SMN gene, and SMN2, the centromeric copy, differ by only 5 base pairs, and the coding sequence differs by a single nucleotide. The 5 main subtypes of SMA are classified according to the International SMA Consortium (ISMAC) system, based on age of onset and maximum motor function achieved. The most common types are acute infantile (SMA Type I, or Werdnig-Hoffmann disease), chronic infantile (SMA Type II or Dubowitz disease), chronic juvenile (SMA Type III, or Kugelberg-Welander disease), and adult onset (SMA Type IV). SMA type 0 has a prenatal onset. SMA type I has onset in the first 6 months of life. SMA type 1 is the most common and severe form, representing 45%





# Standardization of the Bayley scales of Infant and toddler Development for Iranian children



**Abstract**: Objective: The aim of this study was to standardization of the Bayley scales of infant development in 1- 42 months old Iranian children. Materials & methods: The Bayley was performed on 1744 children aged 0-42 months. The normative information was based on a national sample representative of the Iran population for infants 1 through 42 months old in Iran Statistical Center. For each age group, the total raw scores of each scale including; cognitive, receptive and expressive communication, fine and gross motor, were converted to scaled scores with a mean of 10 and a standard deviation (SD) of 3. The composite scores (cognitive, language and motor) derived from sums of scales scaled scores. The composite scores are scaled to a metric with a mean of 100 and a SD of 15, and range from 40-160. Growth scores (range from 200-800, with a mean of 500 and a SD of 100), percentile ranks (range from 1-99, with 50 as the mean and median) and developmental age equivalent determined. To compare the development level of the US children (norm samples) and Iranian children, their mean raw scores in five subscales were compared for finding the difference in scores. Results: The number of girls was 908 (52.1%). Comparing the level of development of Iranian children with the US sample, the mean scores in 28 age groups were different (P value





# The Agreement Between Long- Term Monitoring by Electroencephalography and Magnetic Resonance Imaging in Pediatric Seizure

Fatemeh Zamani <sup>1</sup> © P, Elahe Tari <sup>2</sup>, Morteza Heidari <sup>3</sup>

Sciences, Tehran, Iran

Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran

Abstract: Background: Demonstration of high agreement between structural abnormalities identified on magnetic resonance imaging (MRI) and physiologic abnormalities identified on electroencephalography (EEG) could assist in the assessment of onset zone and etiology of childhood seizure. The present study aimed to assess the agreement between abnormal findings on brain MRI and long-term monitoring (LTM) by EEG as the standard protocol in children with abnormal focal epileptic discharges in LTM. Methods: This cross-sectional study was performed on 95 consecutive children who suffered from seizures with evidence of focal epileptic discharges in LTM which referred to the Children's Medical Center in 2017. All patients were also concurrently evaluated by MRI. Results: Overall, 57 out of 95 patients with abnormal LTM had concurrently abnormal MRI findings. The diagnostic agreement between the MRI and LTM in discovering abnormal findings was found to be high (86.4%) with a kappa agreement coefficient equal to 0.79. Conclusion: About two-thirds of patients with abnormal LTM findings had concurrently abnormal MRI features with high agreement between the two modalities. Thus, MRI and EEG can be valuable in predicting disease cause and prognosis of childhood seizure. Keywords: seizure, magnetic resonance imaging, electroencephalography





# **Top 10 Dysmorphic Syndromes**

# Alireza Biglari, MD PhD <sup>1</sup> © P

Abstract: Top 10 Dysmorphic Syndromes Alireza Biglari, MD PhD Children's Medical Center, Tehran University of Medical Sciences, Tehran, IRAN A dysmorphic neonate causes concern and anxiety for the parents and the physician, so quick diagnosis usually becomes a priority. Establishing a clinical diagnosis, in addition to eliminating uncertainty, allows the estimation of prognosis and interventions that may predict or treat complications, as well as a more accurate assessment of recurrence risk. Although prompt diagnosis has many advantages, again, a hurried but inaccurate diagnosis may have many irreparable consequences. Accurate evaluation of a dysmorphic child depends on good clinical skills, knowledge of phenotypic variations, familiarity with dysmorphology databases and the types of genetic testing. Distinguishing the patterns of dysmorphic features is an essential part of a diagnostic process, as many disorders present with a common set of features. It is obvious that the number of dysmorphic syndromes is so large that it is usually impossible to remember the distinctive features of all of them, and this makes the diagnosis more complicated, especially in the case of rare syndromes. The noteworthy point is that a relatively limited number of these disorders are more common and are visited more frequently by neonatologists and pediatricians. Therefore, knowledge of the special features of common dysmorphic syndromes and their differential diagnoses, as well as sufficient proficiency in using appropriate diagnostic tools, not only leads to timely and accurate diagnosis of common cases but also improves the skill of physicians in diagnosing less common dysmorphic syndromes.





## **Update on SMA management**

Gholamreza Zamani <sup>1</sup> © P

**Abstract**: Spinal muscular atrophy is the second common autosomal recessive neuromuscular disorder due to homozygous deletion or mutation of the SMN1 gene. A nearly identical copy of the SMN1, named SMN2 gene mainly modulates disease severity. The SMA phenotype is majorly influenced by the number of SMN2 copies which usually varies between 1 to 4 and rarely reaches up to 8 copies. The functional difference between both genes is a silent mutation that disrupts exon splicing process through skipping of exon 7 in most of SMN2 transcripts and reducing the functional product of SMN2 to only 10%. With an incidence of about 1:10,000 and high carrier rate of 35-50 in population, SMA is one of the most frequent autosomal recessive disorders in humans. The clinical features of the disease are basically due to the progressive loss of alpha motor neurons in the anterior horns of the spinal cord, which leads to symmetrical atrophy of the proximal muscles of limbs and eventually of the entire trunk. SMA is highly variable disease, 4 major clinical groups depending on the age of onset and the maximum achieved motor abilities are defined. Until 2016 management of SMA was limited to supportive care providing adequate nutrition, respiratory support, orthopedic care and prevention of complications . with introduction of novel therapies now two antisense oligonucleotide formula (Nusinersen, risdiplam) and a vector based gene replacement therapy agent (Zolgensma) are available. Development of these novel therapies for SMA is an exceptional challenge for the scientific community. Meanwhile, SMA is now one of the first treatable inherited diseases. Key words: SMA ,Genetic, Diagnosis, treatment





# **Updates in Pediatric Seizures and Epilepsy**

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Abstract: Abstract-During the last decade there were tremendous changes in the classification, diagnostic as well as therapeutic approaches to epilepsy, epileptic syndromes and epilepsy in adults and pediatric age group. New way of reaching the consensus through qualitative methods such as Delphi and gathering opinion from international experts globally was the corner stone of this new classification. On the other hand by sequencing 90% of human genome through human genome project in 2003, new horizons have been developed in the etiologic diagnosis of epileptic disorders in children and new Paradigms born in diagnostic approach to epilepsy and epileptic syndromes especially in the childhood period. Although we are still at the beginning of genetic engineering and intervention era of treatment and prevention of epilepsy and epileptic disorders in children but personalized medicine and patient care are new effective approaches to treating these disorders and changed old ideas and attitudes regarding treatment of these disorders. In our panel, we will address the abovementioned issues by national experts in the field in an interactive method with participants and audience. Keywords: Epilepsy, epilepsy syndromes, epileptic disorders, childhood, genetic approach, genetic diagnosis and personalized medicine.





# pediatric respiratory distress

مهسا صوتی خیابانی  $^1$  ©  $^0$ 

هیات علمی دانشگاه علوم پزشکی تهران ۱

**Abstract**: The term respiratory distress is used to indicate signs and symptoms of abnormal respiratory pattern. A child with nasal flaring, tachypnea, chest wall retractions, stridor, grunting, dyspnea, and wheezing has respiratory distress. A careful physical examination must be performed when managing a child in respiratory distress To localize site of pathology. The state of responsiveness is another crucial sign. Lethargy, disinterest in surroundings, and poor cry are suggestive of exhaustion, hypercarbia, and impending respiratory failure. Inspiratory stridor, suprasternal, chest wall, and subcostal retractions; and prolongation of inspiration are hallmarks of extrathoracic airway obstruction. whereas expiratory wheezing results from airway obstruction below the thoracic inlet. Grunting is most commonly heard in diseases with decreased functional residual capacity (e.g., pneumonia, pulmonary edema) and peripheral airway obstruction (e.g., bronchiolitis). Typical manifestations of alveolar interstitial pathology are rapid, shallow respirations, chest wall retractions, and grunting. Although respiratory distress most frequently results from diseases of lungs, airways, and chest wall, pathology in other organ systems can manifest as respiratory distress and lead to misdiagnosis and inappropriate management. Respiratory distress resulting from heart failure or diabetic ketoacidosis may be misdiagnosed as asthma and improperly treated with albuterol, resulting in worsened hemodynamic state or ketoacidosis. Careful history and physical examination provide essential clues in avoiding misdiagnosis .Central nervous system (CNS) dysfunction can lead to alterations in respiratory patterns and manifest as respiratory distress. Sepsis and septic shock may cause an acute respiratory distress syndrome (ARDS) with hypovolemic stimulation of baroreceptors, cytokine stimulation of respiratory centers, and lactic acidosis. Other indirect causes of lung injury include systemic inflammatory conditions, trauma, transfusion-related acute lung injury, and pancreatitis. Similarly, renal disease may manifest as respiratory distress by causing metabolic acidosis (e.g., renal tubular acidosis or renal failure).





# A brief review on nutritional support in pediatric intensive care unit: when and how to provide nutrition in a critically ill child?

# Mehrnaz Olfat <sup>1</sup> © P, Masoud Mohammadpour <sup>2</sup>

**Abstract**: In the course of acute illness, many interactions are happened to compensate the initial insult, which is usually associated with compromised energy delivery. These metabolic disturbances usually are characterized by catabolism exceeding anabolism. Although these changes are initially effective, they can lead to severe malnutrition and subsequent increased rate of infection, length of stay in hospital and pediatric intensive care unit (PICU), and morbidity and mortality. Nutritional support, either enteral or parenteral, refer to provision of macronutrient, micronutrient, fluid and electrolyte. The preferred rout of nutrition is enteral feeding. If possible, the most natural route of intake is oral. However, in PICUs, we usually have to provide enteral nutrition (EN) by a gastric tube due to several conditions including decreased level of consciousness, deep sedation, or any other barrier. In the course of admission, nutritional condition even might be deteriorated; so, initial and frequent nutritional assessment and timely nutritional support should be considered as an essential part of pediatric critical care. In the recent years, some consensuses had tried to provide guidelines for nutritional support in PICUs, nonetheless many questions remained unanswered and nutritional support is still a challenge among pediatric intensivists. Herein, we aimed to review the latest guidelines and manuscripts to answer when and how to provide nutritional support in a critically ill child.





#### Picu & Emergency

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#### **ABCDEF** bundle

# Somaye Jafraste <sup>1</sup> © P

Abstract: The ABCDEF bundle is an evidence-based guide for clinicians to coordinate multidisciplinary patient care in the intensive care unit (ICU). A Element: Assess, Prevent, and Manage Pain B Element: Both Spontaneous Awakening Trials (SATs) and Spontaneous Breathing Trials (SBTs) C Element: Choice of Analgesia and Sedation D Element: Delirium: Assess, Prevent, and Manage E Element: Early Mobility and Exercise F Element: Family Engagement and Empowerment Assessment of pain is the first step before administering pain relief. The Behavioral Pain Scale (BPS) and the Critical-Care Pain Observation Tool (CPOT) are the most valid and reliable behavioral pain scales for ICU patients unable to communicate. Coordination of Spontaneous Awakening Trials (SAT) with Spontaneous Breathing Trials (SBT) is associated with decreases in sedative use, delirium, time on mechanical ventilation, and ICU and hospital lengths of stay. Delirium monitoring and management is critically important since it is a strong risk factor for increased time on mechanical ventilation, length of ICU and hospital stay, cost of hospitalization, long term cognitive impairment, and mortality. Early mobility is the only currently known intervention associated with a decrease in delirium duration. Physical therapy is safe and feasible in the ICU, even while on mechanical ventilation, renal replacement therapy, and/or circulatory support.





# Airway management and ventilation methods in basic and advance life support



Abstract: Respiratory problems are the main cause of cardiac arrest in children. In fact, most children and infants who need resuscitation have breathing problems that progress to cardiovascular failure. Airway management, rapid diagnosis and effective treatment of respiratory problems is the basis of PALS (Pediatric Advanced Life Support). In children, lung dysfunction may progress very quickly. So there is little time to waste. The result can be improved by early identification and quick and timely treatment of respiratory problems. Therefore, after identifying the symptoms of respiratory distress and respiratory failure, basic interventions should be performed to support or restore adequate or appropriate oxygenation and ventilation, which requires knowing the types and severity of respiratory distress in children. The provider must intervene quickly to restore adequate respiratory function. Also, he should acquire sufficient skills in basic airway management, including effective ventilation with bags and masks and know the necessary equipment for airway management and choose and prepare the appropriate equipment.





#### Altered mental status in children

#### Golnaz Ghazizadeh Esslami <sup>1</sup> © P

**Abstract**: Acute changes in consciousness vary in degree from mild lethargy and confusion to deep coma. In childhood, the most common causes of coma are toxins, infections, head trauma, hypoxia-ischemia (cardiac arrest, near-drowning), and seizures (postictal state, subclinical status epilepticus.) Disturbances of blood chemistries (glucose, sodium, calcium, bicarbonate, blood urea nitrogen, ammonia) may also produce depressed mental status. The most common cause of long-term morbidity in a patient with depressed consciousness is hypoxia; therefore, airway, breathing, and circulation are addressed first. Vital signs, including pulse oximetry, must be assessed. Breathing patterns may provide important clues. A low respiratory rate may be associated with central nervous system (CNS) depressants or increased intracranial pressure (ICP). Tachypnea may be due to hypoxia, metabolic acidosis, or fever, but in more ominous situations may be due to brainstem herniation. Hypotension may result from severe traumatic brain injury (TBI), spinal cord injury or increased ICP. The glucose level should be checked immediately as a treatable cause of altered mental status. Physical examination searches for clues such as unusual odors, needle tracts, trauma, or signs of dehydration or organ system failure. The Glasgow Coma Scale can be used to assess unresponsive patients regarding their best verbal and motor responses and eye opening to stimulation with score ranging from 3-15 points. Papilledema or paralysis of cranial nerves III or VI in a patient with depressed consciousness is strong evidence of elevated ICP, a medical and neurosurgical emergency. Computed tomography (CT) remains the preferred imaging technique in emergency situations. Lumbar puncture in a patient with elevated ICP can result in transtentorial herniation, so neuroimaging must be performed before LP if elevated ICP is suspected, especially if there are any focal neurological deficits on exam. Beyond supportive measures, the etiology of altered mental status determines the treatment.





#### **Basic Life Support**

#### Zeinab Pourhadi <sup>1</sup> © ®

فلوشیپ مراقبت ویژه کودکان -مرکز طبی کودکان -دانشگاه تهران ۱

Abstract: BLS describes how to perform high-quality CPR for a child and infant and demonstrates the importance of early use of an AED for infants and children younger to han eight years. Early recognition and treatment of cardiac arrest improve survival for children and adults. Effective pediatric BLS by trained healthcare providers or lay rescuers is the foundation of successful resuscitation. Cardiac arrest is a condition defined by the absence of pulses. The two types of pediatric cardiac arrest are hypoxic and sudden cardiac arrest. Individual determinants of survival following pediatric cardiac arrest vary according to the setting(Out of hospital versus in hospital) and individual patient factors. Based on an extensive review of clinical and laboratory evidence, the American Heart Association (AHA)has published frequent updates for pediatric BLS. BLS guidelines differ according to the patient's age. These differences are defined as follows:Infants: younger than one year of age and children: one year of age to puberty. The approach to BLS in infants and children is divided into single-rescuer and two or more rescuers. BLS guideline describes how to determine the unresponsiveness patient, get help and activate the emergency medical response system and assess breathing and pulse. Then describes when to initiate CPR and how to perform compressions, assess rhythm, and use AED.





# **Clinical Characteristics and Complications of** Mechanically Ventilated Children in a Pediatric Intensive Care Unit in Iran: Comparing Different Modes

مهرناز الفت  $^{2}$  مهرناز الفت  $^{2}$  مسعود محمدیور

Medical Sciences, Tehran, Iran

**Abstract**: Background: Mechanical ventilation (MV) is among the most common therapeutic modalities in pediatric intensive care units (PICU), which works based on a defined ventilation mode. Nowadays, conventional and alternative modes including adaptive pressure control (APC) and non-APC modes are frequently employed. Although MV can be helpful in many cases, it may cause some complications resulting in significant morbidity and mortality. Objectives: This study aimed to investigate the demographic features and complications of mechanically ventilated children in a PICU in Iran, as well as to compare different ventilation modes. Methods: A retrospective case-control study was conducted in PICUs of children's medical center hospital - a tertiary referral pediatric hospital. Results: Of 66 patients included in this study, 33 patients were treated with APC modes, whereas 33 patients were treated with non-APC modes. The most common indications for intubation were respiratory failure (53%) and loss of consciousness (13.6%). The mean duration for intubation in patients with and without underlying disorder were 11.7 and 5.2 days, respectively (P-value < 0.01). The means of time for intubation in the APC and non-APC groups were 10 and 11.9 days, respectively (Pvalue 0.145). A total of 23 (34.8%) patients had complications, including death, misplacement of the endotracheal tube, atelectasis, unplanned extubation, etc. There was no significant difference between groups regarding the rates of complications, except for atelectasis. Thirteen (19.7%) patients had atelectasis (2 patients in APC group (6%) and 11 patients in non-APC group (33.3%)) (P-value = 0.022). The mortality rate was the same for the both groups (P-value = 1). Conclusions: In sum, the most common indication for intubation was respiratory failure. No significant difference was observed among patients treated with the APC, and non-APC modes in terms of the complications occurred, except for atelectasis which occurred more frequently in the non-APC group. Therefore, it was concluded that there was no difference between conventional and alternative modes of mechanical ventilation in terms of morbidity and mortality.







#### Definition, classification of shock in children

سید عباس حسنی  $^1$  ©  $^0$ 

دانشیار دانشگاه علوم پزشکی تهران ۱

Abstract: Shock is a physiologic state characterized by a significant, systemic reduction in tissue perfusion and then decreased tissue oxygen delivery and producted harmful metabolics ( lactate). shock is classified into the following stages: 1.Compensated shock: During compensated shock, the body's homeostatic mechanisms rapidly compensate for diminished perfusion, and systolic blood pressure is in the normal range. Heart rate is initially increased. Signs of peripheral vasoconstriction (eg. cool skin, decreased peripheral pulses, and oliguria). 2. Hypotensive shock: For patients with hypotensive shock, The heart rate is markedly elevated, and hypotension develops. Signs and symptoms of organ dysfunction (eg, altered mental status as the result of poor brain perfusion) appear. Systolic blood pressure falls.3.Irreversible shock: During this stage, progressive end-organ dysfunction. Tachycardia may be replaced by bradycardia, and blood pressure becomes very low. In addition to these stages of shock, four mechanisms of shock are recognized: hypovolemic, distributive, cardiogenic, and obstructive:Hypovolemic: Decreased preload caused by volume loss including hemorrhage, gastrointestinal losses, insensible losses (eg. burns), or third spacing.Distributive: Decreased vascular resistance due to vasodilation caused by conditions such as sepsis, anaphylaxis, or acute injury to the spinal cord or brain. Cardiogenic: Decreased cardiac contractility such as primary myocardial injury, arrhythmias, cardiomyopathy, myocarditis, congenital heart disease with heart failure, sepsis,or poisoning. Obstructive :Increased vascular resistance such as congenital heart disease with ductal-dependent or acquired obstructive conditions (eg, pneumothorax, cardiac tamponade, or massive pulmonary embolism), a patient may have more than one type of shock.





#### delirium in picu



Abstract: Pediatric delirium Delirium is a syndrome of acute brain dysfunction. Delirium often manifests with a fluctuating course of severity. delirium is categorized based on psychomotor symptoms. Patient with hypoactive delirium may appear apathetic, withdrawn from environment, or with depressed levels of arousal. Patient with hyperactive delirium suffer from agitation, emotional lability or disruptive behavior, although this is the least common form of ICU delirium. Patient who demonstrates hyperactive and hypoactive behavior having a mixed subtype of delirium. Delirium prevention and management in the PICU: 1-Nonpharmacologic strategies: optimization of sleep hygiene, use of interdisciplinary rounds, family engagement on rounds, and family involvement with direct patient care. There is recommendation to Minimizing benzodiazepine – based sedation in critically ill pediatric patient to decrease rate, duration or severity of delirium. (strong, moderate-level evidence) 2-Pharmacologic management: in critically ill pediatric patients with refractory delirium, haloperidol or atypical antipsychotics can be used for management of severe delirium.





#### Diagnosis and Treatment of Raised Intracranial Pressure

Keyvan Tayebi Meybodi <sup>1</sup> © P, Zohreh Habibi <sup>1</sup>

Abstract: A multitude of neurological and neurosurgical conditions are associated with disordered physiology of the cerebrospinal fluid (CSF) and intracranial pressure (ICP). The position of the brain within skull, creates a unique physiologic environment in which changes in absolute intracranial volume are limited, and thus create pathologic elevations in ICP or shifts in the relative volumetric proportion of intracranial contents. The basic physiologic tenets of this concept were put forward in the Monro-Kellie doctrine. They stated that intracranial volume is composed of V-CSF, V-BLOOD, V-BRAIN, and pathologic items such as tumors or blood clots (V-OTHER), and, this volume must be constant for a given individual. This equation provides a framework for understanding the pathologic causes of elevated ICP and also its treatments. ICP is functionally reflective of the balance among CSF formation, volume storage or compliance, and fluid absorption. Isolated elevations in ICP reduce cerebral perfusion pressure (CPP), which in turn can affect autoregulation and may limit tissue blood flow or even trigger a hyperemic response. Ischemia resulting from raised ICP and reduced CPP causes more swelling, which in turn elevates ICP further. The end point of unchecked ICP elevation is a positive feedback loop terminating in herniation and brain death. Treatments for raised ICP can be grouped according to their site of action within the framework of the Monro-Kellie doctrine. Increased V-CSF can be treated with CSF diversion. Medications such as acetazolamide can reduce CSF production, and their use is a first-line treatment option in chronic conditions such as idiopathic intracranial hypertension. V-BLOOD can be reduced by hyperventilation, head-of-bed elevation, and also sedatives or barbiturates through reducing tissue oxygen utilization. The major pathologic contributor to V-BRAIN is cerebral edema, treatment of which can include hyperosmotics and, in cases of subacute-to-chronic processes, such as tumor-related edema, steroids.





#### **Drowning in children**

# Mahya sadat Mohammadi <sup>1</sup> © P

**Abstract**: Children are at risk of drowning when they are exposed to a water hazard in their environment. The definition of drowning is "the process of experiencing respiratory impairment from submersion/immersion in liquid." The term drowning does not imply the final outcome—death or survival. Use of this terminology should improve consistency in reporting and research; the use of confusing descriptive terms such as "near," "wet," "dry," "secondary," "silent," "passive," and "active" should be abandoned. The injury following a drowning event is hypoxia. Most (71%) drowning deaths in children younger than 1 yr occur in the bathtub, when an infant is left alone or with an older sibling. Infant tub seats or rings may exacerbate the risk by giving caregivers a false sense of security that the child is safe in the tub. The 2nd major peak in drowning death rates occurs in older adolescents, age 15-19 yr. Almost 90% drown in open water. In this age-group particularly, striking disparities in drowning deaths exist in gender and race. Several underlying medical conditions are associated with drowning at all ages. A number of studies have found an increased risk, up to 19-fold, in individuals with epilepsy. Drowning risk for children with seizures is greatest in bathtubs and swimming pools. Cardiac etiologies, including arrhythmias, myocarditis, and prolonged QT syndromes, have been found in some children who die suddenly in the water. Duration of submersion, speed of the rescue, effectiveness of resuscitative efforts, and clinical course determine the outcome in submersion victims. Initial management of drowning victims requires coordinated and experienced prehospital care following the ABCs (airway, breathing, circulation) of emergency resuscitation. The most effective way to decrease the injury burden of drowning is prevention. Drowning is a multifaceted problem, but several evidence based preventive strategies are effective.





#### electrical burns

# الميرا حاجي اسمعيل معمار $^1$ © $^0$

بیمارستان مرکز طبی کودکان ۱

**Abstract**: There are 3 types of electrical burns such as minor(extension cord), high-tension wire, and lightning. Minor electrical burns usually occur as a result of biting on an extension cord. These injuries produce localized burns to the mouth, which usually involve the portions of the upper and lower lips that come in contact with the extension cord. In this category hospital admission is not necessary and care is focused on the area of the injury. The hightension electrical wire burn is the most serious category of electrical burns and need admission for observation. Deep muscle injury is typical and these injuries have a mortality rate of 3-15% for children who arrive at the hospital for treatment. Lightning burns occur when a high-voltage current directly strikes a person or when the current strikes the ground or an adjacent (incontact) object. Lightning burns depend on the current path, the type of clothing worn, the presence of metal and cutaneous moisture. Electrical injuries can involve cardiopulmonary system( cardiac arrest, myocardial ischemia, arrhythmia...), kidney(acute kidney injury, myoglobinuria...) and skin. Also, it has some neurological complications(cerebral edema, mood changes and hemorrhage, seizures....) Electrical burns have some common long-term complications for instance hypertrophic scars, contractures, neuropathic pain. The best way to prevent electrical injuries is to cover all outlets, make sure all wires are properly insulated, tuck wire away from child's reach and provide adult supervision whenever children are in an area with potential electrical hazards.





#### Fluid resuscitation of children shock:

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**Abstract**: The type of shock may not be apparent at presentation. Frequent assessment of the patient's response to fluid resuscitation often provides essential information to guide subsequent treatment. For children with shock, we suggest balanced crystalloid solutions, such as normal saline or Ringer's lactate, rather than colloid. Colloid solutions are more expensive, and patients may develop adverse reactions to them. For children without signs of fluid overload and with hypotensive hypovolemic or distributive shock, the clinician should rapidly administer isotonic crystalloid in a volume of 20 mL/kg and, in patients without improvement, repeat 20 mL/kg fluid boluses as needed up to two or three times over 30 to 60 minutes. For children with signs of fluid overload (eg, rales, worsening respiratory distress, new or worsening oxygen requirement, gallop rhythm, hepatomegaly, or cardiomegaly or pulmonary edema on chest radiograph) or suspected cardiogenic shock, fluid resuscitation should be administered carefully with lower fluid volume over a longer period of time (eg, 5 to 10 mL/kg infused over 15 to 30 minutes). The clinician should have a low threshold for initiating vasoactive medications for persistent shock and endotracheal intubation with mechanical ventilation to treat pulmonary edema in these patients. Excessive fluid resuscitation may be harmful for patients who are not hypovolemic or have compensated shock with Cardiogenic shock, Severe anemia, Severe malnutrition, SIADH, Penetrating injuries to the torso, Obstructive shock caused by tension pneumo- or hemothorax or cardiac tamponade. we recommend that children in resource-limited settings who have severe febrile illness without dehydration or hemorrhage and with normal blood pressures(septic shoc not receive boluses of crystalloid solutions. In a randomized trial of bolus fluid therapy versus no bolus in such patients, administration of 20 to 40 mL/kg of isotonic crystalloid solution or albumin was associated with significantly increased mortality.





#### fluids and medications in PALS

ليلا طاهرنيا  $^1$   $^{\odot}$   $^{\odot}$ 

هیئت علمی ۱

**Abstract**: In Pediatric Advanced Life Support (PALS) Knowing about various resuscitation medications is an important part. These medications are divided into two subcategories: resuscitation fluids and medications. Medication and resuscitation fluids in PALS following appropriate airway management and breathing are inevitable parts to save lives. It is crucial to memorise commonly used drugs in PALS within the field of pediatrics. These primary drugs are a big part of understanding PALS, and the ability to recall PALS drug doses, actions, indications and administrations is our aim to practice in this article.





#### Picu & Emergency

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# **Hemodynamic Monitoring in Shock State**

#### Reza Shabanian <sup>1</sup> © P

**Abstract**: Adequate tissue perfusion and oxygenation in patients with shock are crucial. Hemodynamic monitoring will help to intervene before organ failure or complications occur. It also guarantees the adequacy of the therapeutic measurements in delivering the substrates at the cellular level. There are different types of invasive and noninvasive hemodynamic monitoring including arterial and central venous lines, pulmonary artery catheter, pulse contour cardiac output (PiCCO) measurement, ultrasound cardiac output monitoring (USCOM), near infrared spectroscopy (NIRS) and echocardiography. A combination of hemodynamic parameters can help determine the best individual treatment for critically ill patients.





#### Medical Errors associated with communication failure

فاطمه خانی نوذری  $^2$ , نیلوفر قنبری  $^2$  فاطمه خانی نوذری  $^3$ 

**Abstract**: A medical error is a preventable adverse effect of medical care, whether or not it is evident or harmful to the patient. High error rates with serious consequences are most likely to occur in intensive care units, operating rooms, and emergency departments. Medical errors are also associated with extremes of age, new procedures, urgency, and the severity of the medical condition being treated. (1) Communication issues among providers led to medical errors, and these included: failure to communicate, failure to review the medical record, poor professional rapport, and communication between providers and the patient/family including not using qualified interpreters when applicable. (1) Errors in verbal communication are a common source of medical error. Risk factors for verbal errors include: • Disruptive behavior including rude language or verbal abuse. • Environmental noise issues such as cell phones, pagers, and phones. • Cultural differences among patients and providers. • Hierarchy issues. • Providers acting as autonomous agents. • Personality differences. • Language barriers. • Lack of working as a team. • Multiple conversations are occurring simultaneously. • Socioeconomic variables, such as education and literacy.(2( Communication breakdowns among clinicians, patients, and family members can lead to medical errors, yet effective communication may prevent such mistakes.(3) 1. Carver N, Gupta V, Hipskind JE. Medical Error. [Updated 2022 Jul 4]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK430763/2. Rodziewicz TL, Houseman B, Hipskind JE. Medical Error Reduction and Prevention. 2022 May 1. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. PMID: 29763131. 3.Street RL, Petrocelli JV, Amroze A, et al. How Communication "Failed" or "Saved the Day": Counterfactual Accounts Medical Errors. Journal of Patient Experience. 2020;7(6):1247-1254. doi:10.1177/2374373520925270





## Mild traumatic brain injury in children

کیوان طیبی میبدی  $^1$   $^0$  زهره حبیبی  $^1$ 

دانشگاه علوم پزشکی تهران ۱

Abstract: Traumatic head/brain injury (TBI) is a leading cause of life-long disability in children. Fortunately, the majority of children (90%) suffer only from minor injuries and can be released home after triage in the emergency department. Fall is the most common cause. The male to female ratio for hospitalized patients is 3:2, and boys have four-fold higher risk of fatal trauma. TBI in children differs significantly from adults in multiple aspects, as clinical assessment is frequently challenging, and details about the neurological symptoms at the time of injury may be lacking. Type of trauma varies with the child's age and physical activity. The age-specific biomechanical properties of the pediatric skull, face, brain, and neck muscles make children susceptible for distinctive types of injuries. Mild traumatic brain injury (mTBI) is defined as Glasgow Coma Scale of 14 or greater, and loss of consciousness not exceeding few seconds. Physical symptoms include headache, nausea, vomiting, dizziness, balance problems, vision problems, photophobia, emotional lability, irritability, difficulty remembering, and feeling mentally slow and foggy. Minor head trauma is a subgroup with GCS 15 and normal examination. Over the past decade, the incidence of diagnosed pediatric mTBI has increased exponentially, likely due to increased awareness and recognition. CT scan should be used when the clinician has suspicion for skull fracture or intracranial hemorrhage. The indications for requesting brain computed tomography (CT) include; Suspicion of child abuse, focal neurologic findings, signs of skull fracture, altered mental status, loss of consciousness if longer than a few seconds, bulging fontanelle, persistent vomiting of more than three times, seizure following injury, and high-risk mechanism including fall from a height more than 0.9 m; head struck by high impact object; motor vehicle collision with patient ejection; death of another passenger.





## Non-pharmacologic Management of Pain in Children

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**Abstract**: Non-pharmacologic measures are useful in reducing stress and anxiety in children undergoing invasive procedures. In clinical trials evaluating various psychological interventions for needle-related procedural pain in children, distraction and hypnosis significantly reduced pain and stress. Physical measures, such as massage, heat and cold stimulation, and acupuncture. Behavioral measures, such as exercise, relaxation, biofeedback, desensitization, and art and play therapy. Cognitive measures, such as distraction, imagery, hypnosis, and psychotherapy music therapy . music therapy reduce pain, anxiety, medication requirements, and inflammatory markers in addition to improvements in sleep quality and ability to mobilize. In critically ill neonates, music therapy is also associated with decreased pain during heel prick procedures. A systematic review of music therapy via prerecorded music in postoperative patients, including critically ill children, reported significant reductions in pain scores, anxiety scores, and opioid use in the immediate postoperative period. nonnutritive sucking with oral sucrose be offered to neonates and infants (< 12 mo old) prior to performing invasive procedures strong, high-level evidence) The use of oral sucrose has had the biggest effect in some studies, but combinations of all nonnutritive suck techniques with swaddling have shown additive effects on pain reduction during heel stick procedures with no AEs reported among infants. The use of oral sucrose solution alone is beneficial during painful procedures without evidence of blunting of the analgesic.





# Opoid and amphetamine poisoning in children

سارا معماریان Sara Memarian <sup>1</sup> © ®

Abstract: Poisoning in children is a prevalent and life threatening emergency. Managing this problem needs careful attention to the signs, symptoms and the history that the care giver provide to the medical team. Nowadays narcotic and illegal drugs such as opioids and amphetamines have become abundant all around and even children may be brought to the emergency department by getting poisoned with both of them. opioids mainly have suppressive effects on central nervous and hemodynamic system and the affected child is brought with the presentation of lethargy, drowsiness and bradypnea. On the other hand amphetamines are stimulant and the affected child may show, irritability, hypertension and tachycardia. There is belief among the drug abusers that these two type of drugs have antagonistics effect on eachother and some parents might give amphetamine or opioid to the intoxicated child to neutralize the other drug. The child protection organizations should be more empowered and strong to protect the children of the affected families and this is a big challenge of the health team confronting the poisoned child.





### Pain and Sedation assessment scales

### Zeinab Pourhadi <sup>1</sup> © ®

فلوشیپ مراقبت ویژه کودکان -مرکز طبی کودکان -دانشگاه تهران ۱

**Abstract**: Effective acute, procedural and chronic pain management is critical in PICU.The goal of analgesic therapy is to provide comfort, reduce physiological stress response, risk of addiction, hemodynamic instability and end organ injury. In order to titrate analgesic therapy to effect ,manage pain adequately, and monitor for signs of medication toxicity or adverse effects, pain assessment is of utmost importance. Self-assessment to report pain scores although is gold standard for monitoring the efficacy of analgesic therapy but is largely not possible for the majority of PICU patients. Physiologic indicators and pain-related behaviors(verbal and nonverbal) are unreliable for pain assessment. Behavioral observation scales are the standard of care for children under three years of age. Children ages 4 to 8 years are usually able to selfreport pain. Older children can typically give a self-assessment of their pain using more validated methods such as verbal rating scale, numeric rating scale, and visual analog scale. The Wong-Baker FACES scale and the Bieri faces pain scale Revised are suitable for children of any age and developmental stage. Sedation in the PICU setting is challenging. The optimal condition for the non-neuromuscularly blocked patient would be that she is easily arousable or conscious, comfortable, breathing in sync with the ventilator, and in a state referred to as the Goldilocks Zone(not too deep, not too light). The state behavioral scale, the COMFORT scale, the COMFORT behavioral scale, and the Richmond agitation sedation scale have been validated in PICU patients.

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#### Picu & Emergency

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## Postoperative nutrition in children

Behdad Gharib بهداد قریب <sup>1</sup> © ®

**Abstract**: The nutritional need of the children after surgery is a challenging issue. The exact nutritional requirements are not known. Some authorities believe that because of high level of stress hormones and their counter regulatory effects, there would insulin resistance and the provided nutrients can be consumed by the metabolic system, in the postoperative situation. The CRP level has been proposed as an indicator for increasing the provided energy, the lower CPR level, the more energy and nutrients can be consumed by body. Different diseases have various effects on the metabolism and knowing the pathophysiology of the disease can be proper guide in providing the right amount of nutrient to the patient.





#### Picu & Emergency

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## toilet training

## الميرا حاجي اسمعيل معمار $^1$ © $^0$

بیمارستان مرکز طبی کودکان ۱

**Abstract**: Toilet training should begin when the child is developmentally ready or shows signs of readiness. Pediatric health care providers must be able to recognize and understand the importance of readiness for both the caregiver and the child. They should discuss toilet training with caregivers at each health supervision visit beginning at age 12 months. The child's readiness is based upon the attainment of certain physiologic, developmental and behavioral milestones rather than chronologic age. Caregivers must be prepared for the toilet training process before they begin. They should know how to tell when their child is ready to begin training and should have expectations about the duration of training, accidents, and setbacks. Caregivers should plan toilet training when at least one caregiver is able to devote the time and emotional energy necessary to be consistent on a daily basis for a minimum of three months. Toilet training involves many steps: communicating the need to go, undressing, eliminating, wiping, dressing, flushing, and hand washing. Going through these steps consistently reinforces proper toileting skills. Toilet training is a challenging process that is frequently accompanied by problems and setbacks. Temporary setbacks are a normal part of the process and do not constitute failure; they are expected in times of acute illness, a family move, new child care arrangements, or a family crises. Nocturnal enuresis and toileting refusal are the most common problems in healthy children.





## use of Inotropes and vasopressors in shock

ليلا طاهرنيا  $^1$   $^{\odot}$ 

هیئت علمی دانشگاه تهران ۱

Abstract: Inotropes are one of the most commonly used drugs in intensive care setting. Inotropes are able to boost myocardial contractility and affect on peripheral vascular resistance. In pediatric field, inotropic agents used as first line are norepinephrine and epinephrine. The vasoactive agents indicated in the treatment of shock in children include vasoconstrictors (e.g. norepinephrine, epinephrine and dopamin) or vasodilators (e.g. dobutamine, milrinone). The inotropes selected in treatment of shock are chosen based on perfusion and hemodynamic state, blood pressure, systemic vascular resistance, the end-diastolic volume and cardiac contractility. This essay aims to guide appropriate use of these agents based on their mechanism, effects and proper selection of them according to pathophysiology.

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## Multisystemic Inflammatory Syndrome in Neonates (MIS-N) a review of articles

## Dr Roya Taheritafti <sup>1</sup> © P

**Abstract**: Definition: There is a clear definition of MIS-C in children as described by the World Health Organization (WHO). Overall, the MIS-C definition includes fever with multi-organ involvement (2 or more organs) and evidence of SARS-CoV-2 infection. Currently, there is no agreed definition of MIS-N but we can categorize neonates as having confirmed MIS-N when they fulfilled either the WHO criteria for MIS-C and had a confirmed infection or exposure to SARS-CoV-2 infection before 28 days of age after ruling out other causes. Multisystem inflammatory syndrome in neonates (MIS-N) takes various forms with multiple cardiac presentations and complications, and the treatment methods include various immunological treatments. MIS-N is developed because of immune-mediated multisystem injury either due to the transplacental transfer of maternal SARS-Co2 antibodies or due to late response to antibodies produced by the newborn to SARS-CoV-2 infection. Newly neonates born to mothers with SARS-CoV-2 infection during pregnancy demonstrated evidence of a multisystem inflammatory syndrome with raised inflammatory markers and multiorgan failure that has been described as multisystem inflammatory syndrome in neonates (MIS-N). Clinical manifestations: Regarding clinical features, only 18.2% of MIS-N neonates presented with fever and the cardiovascular dysfunction and respiratory distress are the most common findings in neonates with MIS-N. Gastrointestinal symptoms, mucocutaneous abnormalities, neurological impairment and acute kidney injury were seen in many of MIS-N neonates. Laboratory tests: Laboratory tests showed mostly an increase in C-reactive protein (CRP), Procalcitonin, brain natriuretic peptide (NT-proBNP), troponin, D-dimer, interleukin-6 and thrombocytopenia, metabolic acidosis, prolonged prothrombin time, hypoalbuminemia. Imagines: Chest X-ray and echocardiography are useful for confirmation of diagnosis. Treatments: -Intravenous immunoglobulins (IVIG) - Intravenous steroids (mostly methylprednisolone). -inotropic supports. -Broad-spectrum of antibiotics Conclusion: Neonates born to SARS-CoV-2-infected mothers at any time during pregnancy or during 28 days after birth should be closely monitored for MIS-N.

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#### Neonatology

oral

## **Common Newborn orthopedic problems:**

محمدرضا زرکش $^1$  ©  $^{ ext{P}}$ 

Abstract: Common Newborn orthopedic problems: Newborn orthopedic problems are cause for major concern for the family. Dysplasia of the hip and abnormal feet can be successfully diagnosed and treated with early recognition. Hip Developmental dysplasia of the hip (DDH) is the most common neonatal hip disorder. It was initially thought to be congenital in origin but is now recognized as developmental, hence the change in terminology from congenital dysplasia of the hip to DDH. At birth, an involved hip is rarely dislocated; instead, it is dislocatable. Whether the hip stabilizes, subluxates, or ultimately dislocates depends on postnatal factors. Foot Metatarsus adductus is probably the most common neonatal foot problem. It results from in utero positioning, and is bilateral in approximately 50% of neonates. It is also associated with hip dysplasia; careful examination of the hips is necessary. The calcaneovalgus foot is a common physiologic variant. It results from in utero positioning. This condition is manifested by a hyperdorsiflexed foot, with an abducted forefoot and valgus hindfoot. It is usually associated with external tibial torsion. It typically occurs unilaterally. In utero, the plantar surface of the foot lies against the uterine wall, forcing it into a hyperdorsiflexed, abducted, and externally rotated position. This position produces the calcaneovalgus foot and external tibial torsion. Talipes Equinovarus (Clubfoot) is one of the most common pathologic entities affecting the neonatal foot. It is a deformity of the foot and the entire lower leg. It is classified as congenital, teratologic, or positional. The congenital clubfoot is usually an isolated abnormality, whereas the teratologic form is associated with an underlying neuromuscular disorder. Positional clubfoot refers to a normal foot that has been held in an equinovarus position in utero. any child with a clubfoot deformity requires a careful musculoskeletal and neurologic evaluation to search for other abnormalities.





#### Neonatology

oral

## **Community-acquired Covid infection in neonates**

## Sangsari Razieh <sup>1</sup> © P

**Abstract**: On December 31, 2019, China informed the WHO about an outbreak of pneumonia with unknown causes and isolated the novel coronavirus from these patients in January 2020. Iran reported its first confirmed cases in February 2020. During the virus widespread, there was a disaster among adult patients, and there was little information about the pediatric population and neonates. Nonetheless, despite increasing knowledge and awareness about SARSCOV2 and strictly adhering to all the published protocols and guidelines, different variants and successive mutations of this virus continued to infect children, especially neonates. Most neonatal SARSCOV2 infection is transmitted postpartum due to environmental exposure, and vertical transmission, either congenital or intrapartum, consists of 30% of cases. In our experience, we believe that clinical characteristics of Community-acquired neonates may differ from neonates who are admitted due to perinatal infection. This observational cohort study was performed from October 2020 to March 2022 on community-acquired SARSCOV2 infection in neonates admitted to the NICU or neonatal ward. Neonates were eligible for inclusion in the study if their RT-PCR was positive postpartum. We identified a total of 55 neonates as Community-acquired SARSCOV2 positive. The most common presenting symptom was fever with 35 (61%). Necrotizing enterocolitis was seen in 18% of neonates, and 30% of the cohort were preterm. Neutropenia manifested in 34% of cases. Platelet count in all cases was more than 150×103µ/l. Two neonates had bacterial urinary tract co-infections. All neonates were discharged without complications, and there was no mortality among the studied population. Conclusion: Community-acquired cases seem to be milder than the cases that acquire the virus perinatally. None of our neonates required antiviral, anticoagulant, or corticosteroid therapy and all recovered while receiving only supportive treatment, so the role of these medications in managing neonatal SARSCOV2 infection requires further evaluation. The relatively high prevalence





## Congenital central hypoventilation syndrome

دكتر آرش بردبار $^{1}$  ©  $^{0}$ 

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Abstract: Congenital central hypoventilation syndrome is a rare disorder of the autonomic nervous system with an estimated incidence of about 1 in 148,000 to 200,000 live births. Congenital central hypoventilation syndrome (CCHS) is a rare genetic disorder caused by mutations in the Paired-Like Homeobox 2B (PHOX2B) gene. CCHS typically presents in the neonatal period usually shortly after birth. Presentation of CCHS in newborns were first reported in 1970. Some patients may also present in late childhood or adulthood (late-onset CCHS). CCHS classically manifests as hypoventilation which is worsening in non-rapid eye movement (NREM) sleep, unusually it presents with hypoventilation during waking hours except for severe cases and rarely presents with apnea. The main respiratory manifestation is the incapability of breathing regulation in response to abnormality of CO2 and O2 blood concentration. The most recognized symptom of CCHS is the inability to control breathing that varies in severity, resulting in the need for life-long ventilatory support during sleep in some patients or all the time in others. The disease is characterized by central hypoventilation due to abnormally reduced ventilatory responses to hypercapnia and hypoxia and associated manifestations of autonomic dysfunction such as Hirschsprung disease and neural crest tumours (neuroblastoma, ganglioneuroblastoma, and ganglioneuroma). It classically manifests in newborn babies who present with central apneas, hypoxemia, and hypoxentilation that are most severe during sleep resulting in the need for assisted ventilation. While a subset of patients remain asymptomatic until an older age with a diagnosis of later-onset CCHS Adequate ventilation is essential to ensure optimal growth and development of CCHS patients. Ventilation can be managed with a mechanical ventilator via tracheostomy or masks, or using phrenic pacemakers. Monitoring both oxygen saturations and CO2 using end-tidal capnography at home helps ensure adequate ventilation in all conditions (sleep, awake, during illness and growth spurts).





## Efficacy of Fluconazole Prophylaxis on Invasive Candidiasis Infection in Extremely Low Birth Weight Neonates

Amir Naddaf <sup>1</sup> © P, Hosein Dalili <sup>2</sup>, Nikoo Niknafs <sup>2</sup>, Mamak Shariat <sup>3</sup>, Raheleh Moradi <sup>3</sup>, Hasti Charousaei <sup>3</sup>, ElahehAmini <sup>2</sup>, Roksana Moeini <sup>2</sup>, Afsaneh Akhondzadeh <sup>4</sup>

Abstract: Background: Invasive candidiasis infection is one of the main life-threatening problems for extremely low birth weight (ELBW) neonates who are in the neonatal intensive care unit (NICU). Candidiasis can cause mortality, short-term morbidity, and long-term neurodevelopmental outcome in infected infants who survive. Therefore, since several years ago fluconazole prophylaxis has begun for premature newborns who were admitted to NICUs in some parts of the world. Methods: In this retrospective cohort, the population study was all the infants of less than 1,000 gram admitted to Valiasr Hospital during the years 2011-2016. The subjects were divided into two groups of control and intervention. The control group did not receive any fluconazole prophylaxis, while for the test group, intravenous fluconazole was administered. Finally, we compared the incidence of candidiasis between the two groups. Results: Fluconazole was administered to 70 out of 167 neonates. Our findings showed that two infants of the prophylaxis group (2.9%) and two (1.2%) of the non-prophylaxis group were infected with Candida species. The difference between the two groups was not statistically significant (P=0.501). Among the risk factors, bacterial sepsis, the duration of central catheter installation, total parenteral nutrition, meropenem or vancomycin administration, and hospitalization costs were significantly related to the incidence of invasive candidiasis infection. Conclusion: The incidence of candidiasis in our study was 2.39% and fluconazole prophylaxis has not been effective in reducing fungal infections. Consequently, further investigations in larger sample sizes with different study settings and a variety of methodologies are needed to evaluate the efficacy of fluconazole prophylaxis on invasive candidiasis infection in ELBW neonates.





# **Evaluation of Cerebrospinal fluid (CSF) amino acid levels** in neonates with refractory seizures



نویسنده \_ ارائه دهنده ۱

Abstract: Objective: Seizures are the most important and common symptom of a serious neurological disorder in infancy. Amino acids can play a neurotransmitter role in the brain. Due to the fact that seizures are the occurrence of transient signs and symptoms due to over activity or abnormal activity of neurons in the brain, some anticonvulsant drugs can be useful in the treatment of seizures by inhibiting the mechanism of crude amino acid receptors. Therefore, the study of the amount and pattern of CSF amino acids in patients with seizures can be helpful in the treatment and control of seizures in patients. The aim of this study was to investigate the pattern of CSF amino acids in neonates with resistant neonatal seizures. Material and methods: This prospective study was performed on infants 1 to 56 days old who were admitted to the intensive care unit of the Children Medical Center in 2016 with a clinical diagnosis of refractory seizures. Results: 48.2 % of study participants were female. Clonic seizures were observed in 44.4%, subtle in 14.8%, tonic seizures in 7.4%, tonic-clonic seizures in 11.1% and spastic seizures in 22.2% of participants. There was a significant correlation between glutamic acid amino acid level and EEG (P = 0.007). While in other amino acids, no significant relationship was observed between amino acids and EEG. Also, no significant relation was observed between CSF amino acids and type of seizure. Conclusion: Considering that neonatal seizures can be one of the predictive causes of cerebral palsy and delayed brain development in children, early diagnosis of etiology and treatment is very important in the prognosis of these infants. Therefore, it is suggested that further studies be performed with larger sample size. Keywords: Amino acids, seizures, cerebrospinal fluid (CSF)

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## Exome sequencing in the diagnosis of the dysmorphic child

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دانشگاه علوم پزشکی تهران ۱

**Abstract**: Exome sequencing in the diagnosis of the dysmorphic child A dysmorphic neonate is a cause of concern and anxiety for the parents and the physician. Making a clinical diagnosis allows a targeted search for a genetic aetiology in order to correctly delineate the healthcare requirements of the. Cytogenetics and molecular techniques improve our ability to make precise syndrome diagnoses. Eventhough there is a certain degree of urgency in making a diagnosis in a dysmorphic neonate, a snap diagnosis should never be made. Around 4,000 malformation syndromes have now been delineated and many are associated with medical problems. Thus making a specific syndrome diagnosis can influence immediate medical management. A detailed history, a physical examination for detailing the major and minor anomalies, recording the growth, examination of previous records and photographs are complemented by cytogenetics and molecular genetic techniques in achieving a diagnosis. Familiarity with dysmorphology databases and cross referencing the anomalies especially the rarer ones helps in narrowing the differential diagnosis. The recent development of genetic testing, with an estimated detection rate of 40% for Next Generation Sequencing (NGS) and of 10% for microarrays analyses, allows to perform an etiological diagnosis in at least half of the known syndromes.





#### Neonatology

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## Genetic approach in diagnosis of dysmorphic child

Moeinadin Safavi <sup>1</sup> © ®

**Abstract**: Pediatricians and neonatologists often have the unique opportunity to be the first to identify abnormalities in neonates/infants and children. In this lecture, several abnormalities are discussed along with potential related genetic syndromes and the appropriate genetic approach for their diagnosis. Abnormalities and physical features discussed include birth parameters, aplasia cutis congenita, holoprosencephaly, asymmetric crying facies, periauricular tags and pits, cleft lip with or without cleft palate, esophageal atresia/tracheoesophageal fistula, congenital heart diseases, abdominal wall defects and polydactyly.





#### Neonatology

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## How to deal with covid-19 positive neonates?

## Mahbod Kaveh <sup>1</sup> © P

**Abstract**: The recent viral pandemic in Wuhan, Hubei, China has led to the identification of a new species of beta-coronavirus, able to infect humans, the 2019-nCoV, later named SARS-CoV-2. SARSCoV-2 causes a clinical syndrome named COVID-19, which presents with a spectrum of symptoms ranging from mild upper respiratory tract infection to severe pneumonia, with acute respiratory distress syndrome and frequent death. All age groups are susceptible to the infection, but children, especially infants, seem to be partially spared, having a more favorable clinical course than other age groups. There is currently no clear evidence showing vertical transmission and intrauterine SARS-CoV-2 infection in fetuses of women developing COVID-19 pneumonia in late pregnancy, and even if transmission is possible, the SARS-CoV2 positivity of the mother does not require delivery by caesarean section, does not contraindicate the management of the infant in rooming-in and allows breastfeeding. This review provides an overview on the biology of the virus, on the pathogenesis of the infection, with particular attention to pregnancy and neonatal age, on the clinical presentation of infection in newborns and young infants and summarizes the international recommendations currently available on the clinical care of neonates with SARS-CoV2 infection or at risk of catching the virus. The main objective of the review is to provide an update especially focused to the clinical management of COVID-19 infection in the perinatal and neonatal age. Keywords: SARS-CoV-2; COVID-19; pregnant women; pregnancy; neonates; infants

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# Is early preventive caffeine more effective than late preventive caffeine in premature neonates? A clinical trial

## Peymaneh Alizadeh Taheri <sup>1</sup> © P, Negar Sajjadian <sup>2</sup>, Mahboobeh Jabbari <sup>3</sup>

Tehran, Iran

**Abstract**: Background: Advantages of caffeine for the treatment of apnea of prematurity (AOP) have prompted clinicians to use it as a preventive drug even before the occurrence of apnea. Objective: To compare the effect of early preventive caffeine with late preventive caffeine on the occurrence rate of apnea of prematurity, bronchopulmonary dysplasia (BPD) and radiographic changes, necrotizing enterocolitis (NEC), intraventricular hemorrhage (IVH), patent ductus arteriosus (PDA), the need for mechanical ventilation, the length of mechanical ventilation and the length of hospitalization. Materials and method: In this randomized clinical trial study, 90 preterm neonates with the gestational age of 25-35 weeks were divided into 2 groups: group A received caffeine during the first two days of life (early preventive caffeine) while group B received caffeine during the third to the tenth day of life (late preventive caffeine. The occurrence rate of AOP and other outcomes were the primary outcomes. The adverse effects of caffeine in each group were the secondary outcomes. Results: The total occurrence rate of AOP was significantly higher (32.6%) in the late group versus (6.8%) in the early group (p=0.002). The total occurrence rate of BPD was also significantly higher (37%) in the late group versus (18.2%) in the early group (p=0.047). On the other hand, we found a lower need for mechanical ventilation, shorter length of mechanical ventilation, shorter length of hospitalization, and a lower occurrence rate of PDA, NEC, and IVH in the early group that was not significant. No adverse effect of caffeine was observed in each group. Conclusions: Early preventive caffeine administration was associated with a significantly lower occurrence rate of AOP, BPD, and BPD radiologic changes. As other outcomes occurred lesser in the early group that was not significant, future studies with more participants are recommended. Keywords: Preventive caffeine, early, late, premature neonates, outcomes

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#### Neonatology

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## neonatal jaundice

پروانه صادقی مقدم $^1 \ \mathbb{O} \ \mathbb{P}$ 

دانشگاه علوم پزشکی تهران ۱

**Abstract**: All infants should be visually assessed for jaundice at least every 12 hours following delivery until discharge ,serum or transcutaneous bilirubin should be measured as soon as possible for infants noted to be jaundiced





#### Neonatology

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## Premature baby follow-up

ستاره ثاقب  $^1$  ©  $^2$ 

دانشیار دانشگاه تهران ۱

Abstract: Follow-up Care of Preterm Infants Infants who are discharged from NICU require continued comprehensive clinical care and coordination of all subspecialty care, which is provided by the primary care provider clinician. They communicates with the neonatologist and parents when the infant is getting close to being ready for discharge to home. For the family, contact with both the neonatologist and primary care provider decreases their confusion and anxiety. Preterm infants need to be followed up regularly to assess growth and neurodevelopmental outcome and for early stimulation and rehabilitation. Their first visit is 24 to 48 hours after discharge from the hospital, and the timing of subsequent visits will be announced by the neonatologist during the first visit based on the baby's birth weight and underlying problems. Follow up care of preterm infants contains: 1-Medical care issues: Respiratory health, Immunization, Growth and feeding, Sensory issues that need special follow up include vision and hearing. 2- Neurodevelopmental assessment: Neuromotor problems, Cognitive impairments, Emotional and behavioral health, Social communication difficulties.





## resuscitation and stabilization in newborn suspected to Covid 19

مریم ویسی زاده  $^1$  ©  $^2$ 

استادیار دانشکده پزشکی دانشگاه تهران ۱

Abstract: Newborn with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) test results appear to have minimal burden of illness that is directly associated with a viral infection. In population studies, there is a consistent association of SARS-CoV-2. Asymptomatic SARS-CoV-2 infected mother increases the risk for preterm and medically induced preterm birth. Messenger RNA-based coronavirus 2019 in pregnant women lead to maternal antibody production and transplacental transfer of passive immunity to the neonates. There are potential methods of transmission: Intrauterine transmission through occult maternal viremia and hematogenous spread to the fetus through the placenta or through ingestion of viral particles present in AF, the extent of this mechanism appears to be rare with only a few cases report in literature. Intrapartum transmission through contact with maternal-infected secretion, either respiratory droplets or vaginal secretions at birth time. Postnatal transmission through contact with infected secretions from any infected caregivers, who could be parents or medical staffs. Based on the above-mentioned points, protection of newborn and stabilizing in delivery or operating room are very important. Using personal specific equipment (PSE) like eye glasses guan, 3 layers masks during breast feeding and also separational protocols including appropriate distance between mother and newborn, risk of infection will be lowered.





## The Latest Finding in Neonatal Covid

Saeedi Maryam. <sup>1</sup> © P

Abstract: The Latest Finding in Neonatal Covid Saeedi Maryam. Neonatologist. Children's medical center. Tehran University of Medical Science. INTRODUCTION Data on coronavirus disease 2019 (COVID-19) infections in neonates are limited and consist of two categories. Early-onset and Late-Onset Infection. METHODS: In this review, published articles about neonatal COVID-19 are studied. RESULTS: Early-onset neonatal COVID-19 (between 2 and 7 days after birth) is likely caused by intrapartum or more commonly, immediately after birth. Vertical transmission appeared to be uncommon. Neonatal infection was reported in 1% to 3% of births to mothers with COVID-19.(2) Late-Onset Neonatal COVID-19 Infection, The majority of symptomatic infections in neonates are diagnosed beyond 5 to 7 days after birth. Postnatal transmission by neonatal exposure to maternal respiratory secretions or exposure to infected health care workers or household contacts was the major cause. Neonatal MIS-C (MIS-N) has rarely been reported. (2) Nearly two-thirds were asymptomatic. the most common symptoms were tachypnea and fever. (3) Neonates with MIS-N could be critically ill with myocarditis, myocardial dysfunction, coronary aneurysms, disseminated intravascular coagulation (DIC), necrotizing enterocolitis (NEC)-like illness, hypoxemia, and renal failure (2) Laboratory findings included leukocytosis, lymphopenia, thrombocytopenia, and nonspecific findings of elevated inflammatory markers. Chest radiographs abnormalities were seen. ground-glass opacities have been reported with worsening illness. (2) Management for symptomatic neonates was supportive. (2) CONCLUSIONS: Most neonates infected with SARS-CoV-2 are asymptomatic or develop mild illness. Management for symptomatic neonates is supportive, there is no evidence for the use of antiviral medications and steroids 1-Joan Devin, et al. Epidemiology of Neonatal COVID-19 in the United States. Pediatrics: July 2022 2-Deepika Sankaran, et al .COVID-19: A 2021 UpdatePerinatal SARS-CoV-2 Infection and Neonatal. American Academy of Pediatrics. NeoReviews. May 2021 3- Rachel Y, et al. Do Neonates with COVID-19 Infection Get Severely III? COVID-19 Electronic Health Records Neonatology.September 2022





## Vascular anomalies in neonates: When to worry and when to refer?

رضا آزادی  $^1$   $^{\odot}$   $^{\odot}$ 

متخصص پوست کودکان: بیمارستان پوست رازی ۱

**Abstract**: Vascular birthmarks are common in neonates (prevalence: 20-30%). They are classified according to ISSVA into two major subgroups: Vascular tumors and vascular malformations. Infantile hemangiomas are the most common vascular tumor in infancy with a prevalence of 4%. They may cause functional or cosmetic problems depending on their size and location (periorbital area, lip, nose) or may ulcerate to cause significant discomfort. Large infantile hemangiomas (more than 5 cm) with a segmental distribution can be associated with extracutaneous malformations as part of PHACE syndrome or LUMBAR/SACRAL syndrome. Beard infantile hemangioma may be associated with life-threatening airway hemangiomas. Treatment of infantile hemangiomas in their early proliferative stage is crucial for optimal therapeutic outcomes. Vascular malformations are classified according to their most prominent vessel type. They may be isolated or part of specific syndromic conditions with the involvement of other organ systems. Complex vascular malformations are mostly mosaicism due to early somatic mutations. Genetic advances have led to identifying the main pathogenic pathways involved in these conditions. Diffuse capillary malformation with overgrowth, Klippel-Trenaunay syndrome, CLAPO syndrome, CLOVES syndrome, and megalencephaly-capillary malformation syndrome belong to the PIK3CA-related overgrowth syndromes. Complex and/or life-threatening vascular tumors and malformations are extremely rare events but they represent a considerable therapeutic challenge. Accurate diagnosis and appropriate evaluation and management of these conditions, often require multidisciplinary teams. Early recognition of these conditions may improve therapeutic outcomes and avoid severe complications. We will review some clinical clues to recognize these complicated vascular anomalies that benefit from early referral to the vascular anomalies specialty centers.





#### Neonatology

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## Weaning Protocol in BPD in premature neonates

Kayvan Mirnia <sup>1</sup> © P

**Abstract**: Abstract: BPD is an inflammatory lung disease most prevalent in premature neonates. Oxygen treatment and positive pressure ventilation causes abnormal growth and development of alveoli that impairs weaning from ventilator. Method: In order to wean the premature we initially decreased the Fio2 to 30% and increased the PEEP to6-7cmH2O. When Fio2 decreased to less than 30% we decreased the PIP to 12 cmH2O then we decreased the rate to 35 breath per minutes. Then Ti was increased to 0.5 second. The spo2 target was 92-95%. In cases with increasing work of breath the wean process was stopped. Caffeine was loaded and narcotics were off. Result:we experienced a smooth wean without failure. Conclusion: Increasing PEEP and Ti with low rate wit target spo2 of 92-95% increases weaning success in prematures





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## Hemophilia: Causes, types, symptoms, and treatment

Aziz eghbali <sup>1</sup> © P

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Abstract: Hemophilia: Causes, types, symptoms, and treatment Aziz eghbali MD Pediatric Hematologist &oncologist Iran University of Medical Sciences Haemophilia, is a mostly inherited genetic disorder that impairs the body's ability to make blood clots, There are two main types of haemophilia: haemophilia A, which occurs due to low amounts of clotting factor VIII, and haemophilia B, which occurs due to low levels of clotting factor IX. People with more severe haemophilia experience more severe and more frequent bleeds, while people with mild haemophilia usually experience more minor symptoms except after surgery or serious trauma. In cases of moderate haemophilia symptoms are variable which manifest along a spectrum between severe and mild forms. Treatment and prevention of bleeding episodes is done primarily by replacing the missing blood clotting factors. Clotting factors are usually not needed in mild haemophilia In moderate haemophilia clotting factors are typically only needed when bleeding occurs or to prevent bleeding with certain events. In severe haemophilia preventive use is often recommended two or three times a week and may continue for life. Rapid treatment of bleeding episodes decreases damage to the body. Factor replacement can be either isolated from human blood serum, recombinant, or a combination of the two. Some people develop antibodies (inhibitors) against the replacement factors given to them, Desmopressin (DDAVP) may be used in those with mild haemophilia A Tranexamic acid may be given along with clotting factors to prevent breakdown of clots.





## Allogeneic Hematopoietic Stem Cell Transplantation in Pediatrics



**Abstract**: Thomas et al. performed initial allogeneic hematopoietic stem cell transplantation (Allo-HSCT) in a patient with leukemia after receiving radiation and chemotherapy. The field has developed rapidly and spread worldwide afterwards. First, the allo-HSCT indication limited to a few diseases, including acute leukemias, aplastic anemia, and severe combined immunodeficiency. However, the indications has outspread due to increasing HSCT knowledge and improving technologies. Nowadays, allo-HSCT considers as a standard treatment for various diseases such as hematologic malignancies e.g., acute lymphoblastic leukemia, myelodysplastic syndrome, acute myeloblastic leukemia; congenital or acquired hematopoietic disorders, including aplastic anemia, congenital amegakaryocytic thrombocytopenia; primary immune deficiency e.g., severe combined immune deficiency; and osteopetrosis. Furthermore, it is a therapeutic option for inborn errors of metabolisms, including lysosomal storage disease and peroxisome disorders. The disease status, response to previous treatments, and patient performance are factors influencing allo-HSCT decision making. Furthermore, the outcomes are influenced by the patient's age, donor's type, HLA matching, the conditioning and graft versus host disease prophylaxis regimens type. The most transplant-related mortality causes are relapse, graft versus host disease, and infections. It seems that an individualized risk-based approach in selecting patients before allo-HSCT will eventually lead to a better outcome for all patients.





# autologous hematopoietic stem cell transplantation in pediatric patients

هادی متقی پیشه  $^1$   $^{ ext{ }}$ 

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Abstract: Hematopoietic stem cell transplantation (HSCT) has become a well-established therapy for many severe congenital or acquired disorders of the hematopoietic system and for chemo-sensitive, radio-sensitive or immune-sensitive malignancies in children. Today the prognosis for pediatric tumors, has improved. However, patients with metastatic, refractory or recurrent disease have a dismal prognosis and are candidates for aggressive therapies such as autologous HSCT. In pediatrics, the most common autologous HSCT indications are for the treatment of some solid tumors and lymphomas. Autologous HSCT takes advantage of the steep dose-response curve exhibited by many chemotherapeutic agents in the treatment of pediatric solid tumors. Autologous HSCT is based on the concept that escalating doses of chemotherapy are able to kill greater numbers of tumor cells. While neuroblastoma is the only tumor proven to have an increased survival rate with autologous HSCT compared with standard chemotherapy, this treatment is frequently used to treat other high risk solid tumors such as Ewing sarcoma, Rhabdomyosarcoma, Wilms tumor, osteosarcoma, retinoblastoma, brain tumors, Hodgkin and non-Hodgkin lymphoma. Autologous transplants have also been used to reset the immune system in patients with severe autoimmune disorders. The concept of autologous HSCT was initially developed as an aggressive treatment for malignancy. The use of this technique for the treatment of rheumatic diseases is similar. Aggressive therapy (radiation and/or high dose chemotherapy) is given with the intent to destroy the cells that are responsible for the inflammatory disease. In the process, hematologic stem cells are destroyed, and rescue is accomplished by using the patient's own stem cells that were harvested before treatment. The tumor-specific activity and intensity of agents used for autologous regimens have been shown to be important in improving survival. Keywords: Autologous HSCT, Solid tumors





# Blood transfusion reactions: Diagnostic and therapeutic approach

مریم زادسر $^1$  ©  $^2$ 

**Abstract**: Blood transfusion is a safe supportive medical intervention now. Because of the fact that they are biological products, there are some inherent and inevitable risks for transfusion recipients. Alongside, there are some adverse reactions that could be prevented by choosing the strategy of right products for right patient s in the right time in addition to establishment of patient blood management policies. A wide spectrum of adverse transfusion reaction exists and recorded in medical literature. There are categorized based on the time of appearance (acute and delay), pathogenesis (immune, infectious an others) frequency of occurrence (common and rare) and severity of signs and symptoms (sever and mild to moderate). There is some case fatality rate for reactions like acute hemolytic transfusion reaction (which readily arises due to ABO antigen mismatched blood group transfusion and is a preventable reaction by establishing patient identification system and improving laboratory methods to prevent clerical and documentation errors), Transfusion Related Acute Lung Injury (respiratory failure due to immune reaction) and Transfusion Associated Cardiac Overload (heart failure and pulmonary edema due to overload )however these are not very frequent reactions. Besides, there are more frequent less severe reactions such as allergic reactions and Febrile Non Hemolytic Transfusion Reactions, which still are preventable. Other complications like bacterial contamination could cause just a mild febrile reaction or convert to obvious septic reaction with mortality and morbidity. This is also a preventable transfusion reaction. Amongst late onset reactions, delayed hemolytic transfusion reactions are one of the most challenging that is arisen due to the incompatibility of red blood cell antigens other than ABO. TA-GVHD is a rare but fatal delayed reaction which is mostly preventable. Most infectious complications like viral infections (HIV-HBV-HCV-HTLV-CMV-EBV...), parasitic disease (malaria, leishmania, babesia...) and prion disease (nvVCJD) are late onset complication and reliable tracing system is required for detection and correct diagnosis.



## Haploidentical Hematopoietic stem cell transplantation, when and how?

## Hamid Farajifard <sup>1</sup> © P, Maryam Behfar <sup>1</sup>, Amir Ali Hamidieh <sup>1</sup>

**Abstract**: Hematopoietic stem cell transplantation is curative treatment for more than 70 hematologic and non-hematologic diseases. first of in HSCT, patients need HLA full-match donor for HLA-\*A, \*B, \*C, \*DRb1, \*DQB1, \*DPb1 loci as most polymorphic loci in humankind. Any patients have 30 percent chance to find full-match donor in family. In recent years, by development and progress of donor registry bank, patients' chance increased by 70 percent by use of about 40 million unrelated donors. With all this, significant percentage patients can't find full-match donor and because of urgent HSCT and probability of relapse in malignant patients or severe infection, there is no any other choice except haploidentical HSCT that leukemia patients are more at risk. Any kind of HSCTs act as double-edged sword, rejection and GvHD might be happen and those probabilities are more likely in haploidentical HSCT. So choosing the right haploidentical donor will be avoid patients' complication after HSCT. Immunosuppression prophylaxis drugs prepare environmental condition to prevent severe acute GvHD in haploidentical HSCT patients. So immune reconstitution after HSCT must be determined. By now two protocols of haploidentical HSCT contain T Cell Replete (TCR) and T Cell Deplete (TCD) has been defined that both have own cost and benefit. In TCR protocol patients need younger, sex match, ABO match, NIMA mismatch donor and no donor specific antibody in patients. In TCR, the GvHD will be more likely to happen. In TCD protocol, KIR matching is needed. younger, sex match, ABO match, NIMA mismatch donor is preferred and donor specific antibody in patients must be ruled out. More rejection in TCD protocol is expected.





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## Hemolysis in Children

الهام شاهقلی $^{1}$   $^{0}$ 

دانشیار گروه کودکان، دانشگاه علوم پزشکی تهران ۱

Abstract: Hemolysis in children E. Shahgholi, MD, Pediatric Oncologist Associate Professor Bahrami Children Hospital Tehran University of Medical Science Hemolysis is one of the causes of anemia in children. It can be chronic, acute, hereditary or acquired. Extravascular lysis manifests itself with anemia, jaundice, enlarged spleen and tendency to gallstones, whereas the occurrence of fulminant anemia and hemoglobinuria is an important characteristic of intravascular lysis. Many markers are available to detect lysis. The initial markers include complete peripheral blood count, peripheral blood smear reticulocyte, LDH, bilirubin. Coombs test can distinguish the acquired immune hemolytic anemia from other causes of lysis. Serum haptoglobin and hemopexin disappear in intravascular lysis. Peripheral blood examination is the fundamental guidance for the next diagnostic steps. With these markers, hereditary causes such as membranopathy, enzyme deficiency, and hemoglobinopathies can be distinguished from immunological and non-immunological acquired causes.





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## Introduction to transfusion of cellular blood components

### Azita Azarkeivan <sup>1</sup> © P

**Abstract**: Blood transfusion is the process of transferring blood products into a person's circulation intravenously. In modern medical practice commonly use of blood components. These components divided to cellular such as PackedRBC(PRBC)&Platelets and plasma products include FreshFrozen Plasma(FFP)&crayopercipitate . In Cellular components; whole blood transfusion is in few indication such as massive blood transfusion, exchange blood transfusion in newborns&open heart surgery. The most indication for blood transfusion is use of Packed RBC. Transfusion should be based on the patient's clinical condition. Patients with symptomatic anemia should be transfused if they cannot function without treating the anemia. Symptoms of anemia may include fatigue, weakness, dizziness, reduced exercise tolerance, shortness of breath, changes in mental status, muscle cramps, and angina or severe congestive heart failure; acute blood loss of more than 30% of blood volume. In congenital anemia such as thalassemia they need lifelong transfusion, but in sickle cell we just transfused in acute sickle cell crisis(for stroke prevention). Level of transfusion usually is hemoglobin level less than 8mg/dL. Platelet transfusion may be indicated to prevent hemorrhage in patients with thrombocytopenia or platelet function defects. Spontaneous bleeding through intact endothelium does not occur unless the platelet count is no greater than 50,000/mm3. The level of platelet for transfusion: Platelet count 100'000/mm3in patients who need Surgery or unstable condition, and Platelet count 50'000/mm3in patients who need invasive lab.Procedure,Platelet count 50'000/mm3 in patients With active bleeding, Platelet count 20'000/mm3in patients on Chemotherapy or marrow failure with additional hemorrhagic risk. One unit of random donor platelets should increase the platelet count in adults by 5,000-10,000/mm3, but one unit of apheresis platelets should increase the platelet count in adults by 30,000-60,000 /mm3. One apheresis platelet is equivalent to six random donor platelet .Contraindications to platelet transfusion include thrombotic thrombocytopenic purpura and heparin-induced thrombocytopenia.





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## **Patient Blood Management in pediatric**



Blood Transfusion Research Center, High Institute for Research and Education in Transfusion Medicine, Tehran, IranAss

Abstract: Patient Blood Management in Pediatric Azita Chegini, assistant professor, Blood Transfusion Research Center, High Institute for Research and Education in Transfusion Medicine, Tehran, Iran Children are not small adults and need specific evidence-based strategies designed for them. They need management in severe bleeding and blood transfusion to prevent mortality and morbidity. In children, severe bleeding can be explained as blood loss of more than one circulating blood volume (CBV) within 24 hours, blood loss of 50% of CBV within 3 hours, or transfusion of 10% of total blood volume (TBV) every 10 minutes. These patient blood management strategies include awareness of the patient's risk factors and preparation systems for possible bleeding in high-risk situations. The well-defined lethal triad (coagulation, acidosis, and hypothermia) related to the amount of blood transfusion and its appropriate treatment has great importance. Bleeding management in pediatrics should use Goal-directed therapy including promoting hemodynamic stability by monitoring and controlling vital signs, maintaining end-organ perfusion and oxygen delivery, avoiding overtransfusion through the use of point-of-care and laboratory testing, and appropriate utilization of blood components, reducing harm and side effects associated with transfusion, and maintaining mean systolic blood pressure(SBP) at an average of 55 mm Hg in premature neonates and 110 in adolescents. Optimization of preoperative hemoglobin, limiting blood improving hemostasis, reducing bleeding, correcting coagulopathy, incorporating blood-sparing techniques (including restrictive transfusion practices) are fundamentals in patient blood management (PBM) programs, and should be applied to pediatric surgical patients. There are reports association between preoperative anemia as defined by agestratified norms and in-hospital mortality in children undergoing noncardiac surgery. A multidisciplinary Patient Blood Management (PBM) strategy can help to effectively and safely address the demands and strain on the system caused by a patient with a massive hemorrhage. This allows for the safe and effective optimization of blood component usage while reducing unnecessary transfusions. This program can potentially decrease morbidity and mortality for the patient and decrease healthcare costs for the institution.





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## Plasma products, Introduction and indications

دكتر مستانه علائى  $^1$  ©  $^0$ 

نویسنده مسئول و ارائه دهنده ۱

Abstract: PLASMA Plasma is separated from whole blood and frozen for extended preservation. Fresh frozen plasma(FFP) is frozen at -18°C within 8 hours of collection. Plasma frozen within 24 hours after collection is labeled as PF24. Frozen plasma may be stored for up to 1 year at -18°C or lower. Before transfusion, both FFP and FP24 are thawed at 37°C and transfused within 24 hours. CRYOPRECIPITATED ANTIHEMOPHILIC FACTOR(Cryo) Is the cold insoluble portion of plasma remaining after FFP has been thawed at refrigerator temperatures. It contains approximately 50% VIII(at least 80 IU) and 20% to 40% fibrinogen(250 mg) present in the original plasma unit. Cryo also contains von Willebrand factor(vWF) and factor XIII. Cryo was a treatment in hemophilia A before the development of clotting factor concentrates. Currently, cryo is used mainly as a source of fibrinogen. Cryoprecipitate-reduced plasma(cryo poor plasma) Is the supernatant remaining from the production of cryoprecipitate. It is relatively deficient in HMW vWF retaining normal levels of ADAMTS 13. Cryoprecipitate-reduced plasma has been an alternative to FFP for the treatment of patients with TTP. Plasma Transfusion Plasma may be used to replace any plasma protein deficiency. Concentrates, such as factor VIII, albumin, Prothrombin complex concentrates (PCCs), or immunoglobulin, are preferable for replacement of specific deficiencies because these are highly purified and standardized. Plasma is most commonly used either for replacement of coagulation factor deficiencies when no factor concentrate is available or for replacement of multiple factors. In general, a dose of 15 to 30 mL/kg is necessary to achieve a hemostatic effect. Plasma may also be used for rapid reversal of warfarin and is also a source of ADAMTS 13. Cryoprecipitate Transfusion Cryo can be used for factor VIII or vWF replacement and is a good source of fibringen and factor XIII. Some patients may require additional supplementation of fibrinogen using cryo or fibrinogen concentrate.





# Prevalence of microcytic anemia among newborn infants from baharloo hospital, tehran, iran

ندا  $^{3}$ , محمد کاظم بخشنده بالی  $^{2}$ , محمد کاجی یزدی  $^{3}$ , محمد کاظم بخشنده بالی  $^{5}$ , منیرالسادات سهل آبادی  $^{5}$  حکیمیان  $^{6}$  اصغر قربانی  $^{5}$  زهرا ملااسماعیلی  $^{5}$ 

استادیار مغز و اعصاب کودکان گروه بیماریهای کودکان، دانشکده پزشکی بیمارستان بهارلو دانشگاه علوم پزشکی تهران <sup>†</sup> متخصص کودکان و نوزادان گروه بیماریهای کودکان دانشکده پزشکی، دانشگاه علوم پزشکی اصفهان، ایران <sup>†</sup> گروه مامایی،دانشکده پرستاری-مامایی بیمارستان بهارلو، دانشگاه علوم پزشکی تهران، تهران، ایران <sup>۵</sup> استادیار متخصص کودکان و نوزادان گروه بیماریهای کودکان، بیمارستان بهارلو، دانشگاه علوم پزشکی تهران، تهران، ایران <sup>۶</sup>

زمینه و هدف: آنمی میکروسیتیک فقر آهن شایعترین اختلال تغذیهای و از مشکلات بهداشتی عمده در زمینه و نوزادان و کودکان است که با رشد و تکامل ناکافی آنها همراه است. مطالعه حاضر با هدف تعیین شیوع آنمی میکروسیتیک روش بررسی: تعداد .در نوزادان در بدو تولد در بیمارستان بهارلو تهران تابعه دانشگاه علوم پزشکی تهران انجام شد ۲۱۰ نوزادی که بین مهر ماه ۱۳۹۷ تا اسفند ماه سال ۱۳۹۷ در بیمارستان بهارلو تهران بهدنیا آمدهاند مورد بررسی خون بندناف از نوزادان تازه بهدنیا آمده گرفته شد و سلولهای خونی مورد بررسی قرار گرفت. با ۵۰٪ در گرفتند بررسی نتایج شاخصها آنمی میکروسیتیک با شاخصهایی مانند بررسی نتایج شاخصها آنمی میکروسیتیک تشخیص داده شد و بررسی رابطه آنمی میکروسیتیک با شاخصهایی مانند یافته ها: میزان نوزادانی که میزاد و میزان نوزادانی که اطلاعات دموگر افیک صورت گرفت آنمی میکروسیتیک دارند ۹/۰ میباشند. ارتباط معناداری بین سطح غلظت هموگلوبین خون و آنمی میکروسیتیک نشان داده شد. ارتباطی بین سن مادر، وزن نوزادان، قد نوزادان، نوع زایمان، سن حاملگی و نسبت والدین با بروز آنمی میکروسیتیک مشاهده نشد نتیجه گیری: آنمی در نوزادان متولد شده در بیمارستان بهارلو از شیوع نسبتا بالایی برخوردار .میکروسیتیک مشاهده نشد است، بنابراین، امر غربالگری و بررسی بیشتر از نظر ابتلا به بیماری کمخونی خصوصا آنمی میکروسیتیک و عوامل مرتبط با آن از اهمیت بسیار بالایی برخوردار است. نتایج مطالعات نشانگر آن است که آنمی یک بیماری چند فاکتوری میباشد و بروز آن به عوامل مختلف بستگی دارد و نیاز مند غربالگری و تشخیص بهموقع جهت بکارگیری درمان موثرتر میباشد و بروز آن به عوامل مختلف بستگی دارد و نیاز مند غربالگری و تشخیص بهموقع جهت بکارگیری درمان میباشد میباشد و بروز آن به عوامل مختلف بستگی دارد و نیاز مند غربالگری و تشخیص بهموقع جهت بکارگیری درمان میباشد





## **Regenerative Medicine in Pediatric Patients**

Amir Ali Hamidieh  $^1$  ©  $^{\circ}$ , Leila Jafari  $^1$ , Rashin Mohseni  $^{\circ}$ , 00000 00000000  $^{\circ}$ , 000000 00000  $^{\circ}$ , 000000  $^{\circ}$ ,

## Maryam Behfar <sup>1</sup>

**Abstract**: Regenerative Medicine (RM), a novel modality in medical research, is based on a comprehensive knowledge of cell therapy and tissue engineering techniques and gene therapy which aims to replace human cells, tissue or organs in order to rectify erroneous functionality. The era of Regenerative Medicine as future medicine is upon us. Thousands Ideas to market have demonstrated profound, durable and potentially curative effects that are already improving human quality of life for patients who have no other available therapeutic options. Until 2021 products reports in regenerative medicine field included three main branches of gene therapy, cell therapy and tissue engineered based therapy. There are numerous clinical trials in different phases and achieved unprecedented bench to bedside clinical success Stem cells are characterized by their unique pluripotency and self-renewal properties which enables them to differentiate into a variety of specialized cells. The use of Stem Cell Therapies can be categorized into three distinctive eras. In the commencing era, Multipotent Somatic Stem Cells, such as hematopoietic stem cells (HSCs), mesenchymal stem cells (MSCs) and fetal stem cells (FSCs), were utilized. In the subsequent era, pluripotent stem cells including Embryonic Stem Cells (ESCs) and induced Pluripotent Stem Cells (iPSCs) came into play. In the third era, which is the present era, enhanced therapeutic agents including Gene Therapy and Gene Editing have become popularized. Currently, retinal pigment epithelial (RPE) cells derived from ESCs in blindness treatment, pancreatic islet cell therapy in diabetic patients and use of MSCs in GVHD patients after transplantation have created many hopes in RM. Gene therapy including genetic engineering and genetic modification, have provided advanced therapeutics for many disorders such as metabolic and neurological diseases, immune deficiencies and hematological disorders. In addition, related market authorized immunocell therapy products are increasing such as six CAR T cell therapy products approved by

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### What is thalassemia?

## Mohammad kajiyazdi <sup>1</sup> © P

**Abstract**: What is thalassemia? Thalassemia is a hereditary blood disorder in which the amount of hemoglobin and red blood cells in the affected person's body is lower than normal. Thalassemia is a hereditary disease, which means that at least one of your parents must be a carrier of the disease. In fact, thalassemia is caused by genetic mutation or deletion of some specific gene parts. If only one of your parents is a carrier of thalassemia, you may have a form of thalassemia called thalassemia minor. In this case, you will probably not have the symptoms of thalassemia, but you will be a carrier of the disease. Some people with thalassemia minor have mild symptoms. But if both your parents are carriers of thalassemia, you are more likely to develop a more serious form of the disease. The type of thalassemia you have depends on the number of gene mutations you inherit from your parents and which part of the hemoglobin molecule is affected by the mutation. have taken. Thalassemia symptoms may vary depending on its type and severity. Most children with moderate to severe thalassemia show signs and symptoms of thalassemia within the first two years of life. If your child has thalassemia, blood tests may show the following: anemia. Hypochromia, microcytosis, anisocytosis. If you have mild thalassemia, you may not need treatment. But if you have a more severe form of this disease, you may need frequent blood transfusions. There are a series of tests that are done before the baby is born that will determine if your baby has thalassemia and also determine its severity.





## Approach to Microscopic hematuria in children

# Azadeh Afshin <sup>1</sup> © P, Sayed Yousef Mojtahedi <sup>1</sup>, Zahra Noparast <sup>1</sup>

**Abstract**: Definition: Microscopic hematuria is a common finding in children, with 3 to 4 percent of normal school-age children. Microscopic hematuria is defined as the presence of more than five RBCs per high-power field. It may be discovered as an incidental finding on an urinalysis prompted by urinary or other symptoms. The microscopic examination is the gold standard for the detection of microscopic hematuria. Both benign and serious conditions can cause microscopic hematuria in children. It is crucial to identify a potential site of bleeding (glomerular versus nonglomerular) and aid in determining the underlying cause. The presence of red cell casts, proteinuria, and/or dysmorphic RBCs (by an experienced observer) indicates a glomerular source of bleeding. In nonglomerular causes urinary tract infection, idiopathic hypercalciuria and other benign conditions must be excluded. The evaluation depends upon the clinical presentation, which falls into three categories: • Asymptomatic isolated microscopic hematuria • Asymptomatic microscopic hematuria with proteinuria • Symptomatic microscopic hematuria Result: Based on the history, physical examination, and urinalysis, a preliminary diagnosis will be made in the majority of cases and will guide further evaluation and/or intervention. Children with persistent asymptomatic isolated hematuria and a completely normal evaluation should have their blood pressure and urine checked every 3 mo until the hematuria resolves





### **Essentials of Hematuria In Children**

## Sayed Yousef Mojtahedi <sup>1</sup> © P, Zahra Noparast <sup>2</sup>, Azadeh Afshin <sup>3</sup>

University of Medical Sciences, Tehran, Iran

Medical Sciences, Tehran, Iran

**Abstract**: Overview. When blood appears into urine, it's called hematuria. It can be common and is alarming. Many conditions that produce hematuria in children usually are benign ,however there are serious etiologies such as urinary tract tumours and malignancies that can show hematuria initially. Types. There are two types of hematuria: Microscopic hematuria is when there is not enough blood to change the color of the urine and blood can be seen only with a microscope. Gross hematuria is when you can see the blood even without a microscope and urine is red or tea-colored. Causes. There are many different causes of hematuria in children. Some of these causes don't affect the urinary tract, such as menstrual bleeding, vigourous exercise, or infection of the urethra. some causes involve the urinary tract such as tract infections, vesicoureteral reflux, kidney or urinary hypercalciuria,trauma to the urinary tract. Signs and symptoms. Hematuria doesn't have any symptoms other than the identification of red urine ,however, the conditions that cause hematuria may produce symptoms. For example, if the renal stone cause of the hamaturia, pain, dysuria and neusea may be seen. Diagnosis. For diagnosis of hematuria first of all ,urinalysis and urine culture may be obtained.In some cases kidney function tests, electrolytes, urie calcium can complete the lab data. Sonography of urinary tract, CT scan or even MRI may be needed. Tretment. Mostly, hematuria doesn't need any treatment. If it only happens once, it's nothing to worry about. If the r condition is causing the hematuria is diagnosed, it is better to treat that condition. For instance, hematuria due to urinary tract infection (UTI) is treated with proper antibiotics.





## **Etiology of Hematuria in Children**

## Mohsen Akhavan Sepahi <sup>1</sup> © ®

Abstract: Hematuria, defined as the persistent presence of more than 5 RBC/HPF in uncentrifuged urine. Evaluation of the child with hematuria begins with a careful history, physical examination, microscopic urinalysis, family history, identification of anatomic abnormalities, malformation syndromes, hypertension, edema and heart failure. A careful family history is critical in the initial assessment of the child with hematuria given the numerous genetic causes of renal disorders. The most common cause of gross hematuria is bacterial or viral urinary tract infection. Recurrent episodes of gross hematuria suggest IgA nephropathy, Alport syndrome, or thin glomerular basement membrane disease. Hereditary glomerular diseases include hereditary nephritis, thin glomerular basement membrane disease. Hereditary hematuric renal diseases include both autosomal recessive (ARPKD) and autosomal dominant polycystic kidney diseases (ADPKD), atypical hemolytic-uremic syndrome, urolithiasis, and sickle cell disease. Physical examination may suggest possible causes of hematuria. The presence of hypertension (HT), edema, or signs of heart failure suggests acute glomerulonephritis (GN). Hematuria associated with GN is typically painless. Diseases commonly manifesting as GN include postinfectious GN, immunoglobulin A (IgA) nephropathy, membranoproliferative GN, Henochhönlein purpura (HSP) nephritis, systemic lupus erythematosus (SLE) nephritis, granulomatosis with polyangiitis, microscopic polyarteritis nodosa, Goodpasture syndrome, and hemolytic-uremic syndrome (HUS). A history of recent upper respiratory, skin, or gastrointestinal infection suggests postinfectious glomerulonephritis, HUS, or HSP nephritis. Rash and joint complaints suggest HSP or SLE nephritis. Frequency, dysuria, and unexplained fevers suggest a urinary tract infection. Several malformation syndromes are associated with renal disease, including VATER (vertebral body anomalies, anal atresia, tracheoesophageal fistula, and renal dysplasia) syndrome. Abdominal masses may be caused by bladder distention in posterior urethral valves, hydronephrosis in ureteropelvic junction obstruction (UPJO), ADPKD, or Wilms tumor. Hematuria seen in patients with neurologic or cutaneous abnormalities may be the result of a number of syndromic renal disorders, including tuberous sclerosis, von HippelLindau syndrome,





## Evaluation of gross hematuria in children

# Zahra Noparast MD <sup>1</sup> P, Sayed Yousef Mojtahedi MD <sup>2</sup>, Azadeh Afshin MD <sup>1</sup> O

urine that is visible to the naked eye. Red or brown urine can be due to causes other than blood for example from drugs (phenazopyridine, rifampicin....) or foods or metabolites and Free hemoglobin or myoglobin. The aims of evaluation of hematuria are 1) localize the site of bleeding and 2) determine the cause of hematuria. Thus, the first step of evaluation is to establish whether or not the urine discoloration is due to blood. The most etiologies for gross hematuria in children include UTI, irritation of the meatus, nephrolithiasis and trauma. Other less common causes include SCA, coagulopathy, glomerular disease including PSGN and IgA nephropathy, Wilms tumor, and drug-induced hemorrhagic cystitis, Alport syndrome and underlying congenital anomalies. The clinician can establish the underlying etiology by a complete history, physical examination, and urinalysis. Urinalysis may suggest an underlying etiology and potential site of bleeding (glomerular versus nonglomerular). Moe evaluation included CBC, U/C, urine calcium ratio, testing family members for hematuria creatinine, C3, ANA, ultrasonography or CT scan, renal biopsy in some cases and rarely cystoscopy. The majority of symptomatic children who present with gross hematuria have an easily recognizable and apparent cause generally detected by findings of initial evaluation. Asymptomatic hematuria may be also occurred due to Hypercalciuria, UPJO, VUR, nutcracker syndrome and IgA nephropathy.

**Abstract**: Gross hematuria is defined by the presence of an increased number of red blood cells (RBCs) in the urine that is visible to the naked eye. Red or brown urine can be due to causes other than blood for example from drugs or foods or metabolites and precipitated urates. Red urine can occur in Hemoglobinuria, myoglobinuria. Thus, the first step of evaluation is to establish whether or not the urine discoloration is due to blood. The aims of evaluation of hematuria are 1) localize the site of bleeding and 2) determine the cause of hematuria. The clinician can establish the underlying etiology by a complete history, physical examination, and urinalysis. Urinalysis may suggest an underlying etiology and potential site of bleeding (glomerular versus extraglomerular). A glomerular origin of the gross hematuria is usually accompanied by signs and symptoms of fluid overload, high blood pressure, and proteinuria and maybe preceded by history of respiratory or skin infection. Extraglomerular gross hematuria may be also occurred due to hypercalciuria, UPJO, VUR, nutcracker syndrome and underlying congenital anomalies. The most etiologies for gross hematuria in children include UTI, irritation of the meatus, nephrolithiasis and trauma. Other less common causes include SCA, coagulopathy, glomerular disease (PSGN and IgA nephropathy, ...), Wilms tumor, and drug-induced hemorrhagic cystitis. Recurrent gross hematuria can also be seen in children with Alport syndrome and thin GBM disease. In patients who have extraglumerolar hematuria, further evaluation is done by U/C, ultrasonography or other imaging, urine calcium ratio, coagulation test and cystoscopy in some cases. More evaluation to determine glomerular diagnosis are included CBC and smear, urine for casts and protein, BUN, Cr., albumin, electrolytes and other testing driven including C3,C4 , ANA, ANCA, hepatitis B and C , HIV, audiology screen, renal biopsy in some cases.

#### Nephrology

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## **Evaluation of Hypertension in children**

بهناز بازرگانی  $^{1}$  ©  $^{0}$ 

**Abstract**: Hypertension in childhood and adolescence may contribute to premature atherosclerosis and the early development of cardiovascular disease (CVD). As a result, identifying children with HTN and successfully treating their HTN may have an important impact on long-term outcomes of CVD. The diagnosis of persistent childhood HTN is made when repeat BP values on three separate visits are greater than the 95th percentile for the age, sex, and height of the patient, or it is  $\geq 130/80$  mmHg. Stage 1 HTN is defined as SBP and/or DBP between  $\geq 95$ th percentile to





## Fluid therapy in severe malnutrition in children

<sup>1</sup> © P مجتبى فاضل

**Abstract**: Fluid therapy in severe malnutrition in children Malnourished children often present with acute and persistent diarrhea and reduced homeostatic capacity to cope with water and potassium deficits. The pathogens responsible for this diarrhea are fairly similar to those of non-malnourished children in similar settings. Diarrhea and severe dehydration are strongly linked to increased smortality among severely malnourished children, so it is essential to manage these problems optimally. Children with severe acute malnutrition who present with some dehydration or severe dehydration but who are not shocked should be rehydrated slowly, either orally or by nasogastric tube, using oral rehydration solution ORS (5-10 mL/kg/h up to a maximum of 12 h). 2. ReSoMal (or locally prepared ReSoMal using standard WHO lowosmolality oral rehydration solution) should not be given if children are suspected of having cholera or have profuse watery diarrhea (Three or more loose or watery stools in a day, for more than 14 days). Such children should be given standard WHO low-osmolality oral rehydration solution that is normally made, i.e. not further diluted. 3. Any Child with acute diarrhea should go to the oral rehydration center (ORC), in which MUAC screening is ensured, if the child is malnourished, he should be shifted to Dirrhea treatment center (DTC) to be rehydrated, once rehydrated and diarrhea & vomiting improved, he should be referred to OTP or TFC/SC as per his malnutrition condition 4. Child with severe acute malnutrition should be rehydrated by using ORS in the diarrhea treatment centers as per WHO guidelines till he becomes rehydrated, (no use for IV fluids unless it is required as per the guidelines), once rehydrated, the child with SAM should be referred to TFC/SC.





#### Nephrology

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# Management of hypertension in neonates and infants



هیئت علمی دانشگاه علوم پزشکی تهران-مرکز طبی کودکان <sup>۱</sup>

**Abstract**: Data are limited regarding the treatment of hypertension in neonates and older infants. The approach to management is similar to that used in older children with decisions based on the severity of hypertension, the underlying cause, and other clinical factors that impact the well-being of the patient. For asymptomatic neonates and infants with mild hypertension (systolic BP 95th to





## Pharmacologic Therapy in children with hypertension

مستانه مقتدری  $^{1}$  ©  $^{0}$ 

chronic kidney research center r

Abstract: Choice of initial medication in hypertensive children is often confusing. Consideration should be given to special conditions such as underlying CKD or hared disease, or the concurrent presence of diabetes. Some medications may be contraindicated in special circumstances, such as angiotensin converting enzyme inhibitors (ACEI) in patients with bilateral renal artery stenosis. Additionally, if the child or adolescent is found to have a monogenic form of HTN there is often a specific agent that will target the mechanism of the HTN. First-line antihypertensive agents recommended include long-acting calcium channel blockers, ACEI, angiotensin receptor blockers (ARB), and thiazide diuretics. Thiazide diuretics don't use as Initial and single therapy. Other classes of agents including beta- and alphaadrenergic blockers are generally reserved for second- or third-line use given their known side effects. Monotherapy with long-acting drugs that can be dosed once daily is preferred, but additional agents may be needed to achieve optimal BP levels. Children and adolescents have been started on antihypertensive medications should be seen frequently at first, perhaps every 6–8 weeks, so that drugs can be titrated, and then less often once a stable regimen and the goal BP attained. ABPM is a useful tool to help determine response to medication treatment. children treated with specific medication classes – ACEI, ARBs and diuretics – require periodic laboratory monitoring to assess for electrolyte disturbances and other medication related toxicities, and those with known target organ effects such as LVH should undergo periodic reassessments as indicated. Treatment of acute severe HTN requires continuous monitoring of BP to prevent excessively rapid lowering of BP.





## Postoperative electrolyte derangements

آرش عباسی  $^{1}$   $^{0}$ 

هیئت علمی دانشگاه علوم پزشکی تهران-مرکز طبی کودکان ۱

Abstract: Postoperative patients are prone to electrolyte derangements resulting from intravenous fluid therapy, transfusion, stress hormones, nutrition support, refeeding syndrome, acid-base abnormality, fluid loss, and tissue trauma. Excess urinary loss can occur with autodiuresis during recovery from surgery or head trauma. The patient's underlying medical condition prompting the surgery and treatment of other medical conditions may also contribute to electrolyte derangement. Postoperative patients with the following conditions and treatments should have their electrolytes monitored on a daily basis; more frequent monitoring may be indicated in cases of electrolyte deficiency or excess (see 'Laboratory monitoring' above): •Fluid resuscitation •Blood transfusion •Severe organ dysfunction •Traumatic brain injury •Continuous bladder irrigation •Abnormal bodily fluid losses •Large surface area open wounds or burns •Maintenance fluid therapy in patients who are not eating •Postoperative ileus or bowel obstruction •Parenteral nutrition To avoid complications of electrolyte deficiency in the postoperative period (eg, ileus, arrhythmia, seizure), electrolyte deficiencies should be identified and treated. Electrolyte replacement to a goal of normal serum electrolyte levels (as defined for a particular institution) is a safe practice. For the patient with ongoing losses, target the middle of the normal range. Electrolyte deficiencies can be replaced using the intravenous or enteral route. One or the other route of administration may be more appropriate depending upon the clinical situation. Although an enterally administered replacement is preferred for those with gut function, these may be poorly tolerated in postoperative patients. For patients with moderate-to-severe electrolyte deficiency, intravenous administration provides reliable and rapid delivery. The amount and rates of administration are reviewed above. Electrolyte excess in postoperative patients is often related to the underlying disease for which surgery was indicated and its treatment. Enteral options for treatment may not be feasible in postoperative patients





# The importance of performing scintigraphy at the onset of the disease in children with the first episode of febrile UTI

## Neamatollah Ataei <sup>1</sup> © P

مرکز تحقیقات بیماری های مزمن کلیه در کودکان دانشگاه علوم پزشکی تهران ۱

**Abstract**: The importance of performing scintigraphy at the onset of the disease in children with the first episode of febrile UTI Neamatollah Ataei M.D. Pediatric Nephrologist Pediatric Chronic Kidney Disease Research Center Executive Director of the Second Phase of the National CKD Registration in Children Tehran University of Medical Sciences (TUMS) Urinary tract infections are among the most common bacterial infections in children. Renal cortical scintigraphy with technetium-99 m-labeled dimercaptosuccinic acid (DMSA), although not perfect, appears to be the best clinically applicable standard of reference for the diagnosis of acute pyelonephritis (APN). Its sensitivity and specificity for confirming APN are 91% and 99%, respectively, with a total of 97% agreement between scintigraphy and histopathological findings. In 2011, the American Academy of Pediatrics (AAP) recommended that renal and bladder ultrasound (RBUS) be performed after the first febrile Urinary tract infection (UTI). Significant controversy surrounded this recommendation, and many Authors disagreed with the AAP guidelines. In different studies, renal parenchymal involvement on DMSA scan at the onset of infection has been reported between 44.1% and 88% (mean = 67.9%). RBUS findings were in favor of APN in only 30% of cases and normal RBUS was reported in 70% of children despite renal parenchyma involvement. The main limitations of RBUS are the dependence on the equipment and the operator, and the impossibility to obtain data on renal function and very low sensitivity of ultrasound (11% - 40%) in terms of predicting kidney parenchyma damage in APN. The AAP guideline suggest that DMSA scan can be performed after the acute phase of the disease has resolved. In the study of Coulthard et al. in 2014, the findings showed that kidney involvement was missed and scarring occurred in one or both kidneys in 8% of children treated according to NICE guidelines. If, according to the 2011 guidelines, the strategy in assessing children with APN is limited to ultrasound findings, then the identification of a significant number of infected and abnormal kidneys is neglected and will eventually lead to miss patients. Therefore, ultrasound should not be used as the primary imaging technique for the diagnosis of APN.

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## **Evaluation of Delayed Puberty in children**

# Reihaneh Mohsenipour <sup>1</sup> © P

**Abstract**: Delayed puberty is defined as Absence of breast development by age 12 to 13 years in girls or absence of testicular enlargement by age 13 to 14 years in boys. Therefore we should be aware that Onset of pubic hair is not usually included in this definition, because this is typically a sign of adrenarche rather than true puberty. About 5 % of healthy individual in population may experience delayed puberty. In delayed puberty we have hypogonadism in different degree. It is classified in two groups: primary and secondary. Primary hypogonadism is diagnosed with low gonadal steroid and high LH and FSH .Secondary hypogonadism is diagnosed with low gonadal steroid and low to normal LH and FSH. For evaluation, first complete history and physical exam should be done then testing such as CBC, ESR, BUN, cr , LFT, TTG (IgA) needs to rule out underlying condition such as celiac, malnutrition, anorexia nervosa, IBD. At final step hormonal analysis needs to be done. These include LH , FSH , Estradiol or Testosterone , Prolactin , IGF1 , TFT . In some cases further evaluation needs to be done. This includes karyotype, brain MRI, olfactory function testing and genetic testing panel. After definitive diagnose, appropriate treatment will be done. In some case of secondary hypogonadism for differentiation of constitutional delay of growth and puberty with GnRH deficiency a short coarse of hormonal therapy is helpful.





## **Growth in Turner syndrome**

رضا توكلى زاده $^1$   $^{\odot}$ 

**Abstract**: Turner syndrome (TS), first reported as a clinical syndrome (prior to the availability of karyotyping) in 7 women with short stature, sexual immaturity, neck webbing, and cubitus valgus in a paper published in 1938 by Henri Turner, an Oklahoma physician. Prevalence is approximately 1 in 2000-2500 live female births, however many patients with a milder phenotype may remain undiagnosed. Most Genotypes associated with TS are 45,X and 45,X with mosaicism. 10-12 percent of all individuals have mosaicism involving a cell line containing Y chromosome material (Y-chromosome mosaicism), that may be identified if marker chromosomes (sex chromosome material of uncertain origin) are detected on the karyotype or if patients present with virilization. The most consistent characteristic of TS is short stature, present in virtually 100% of patients. Other anomalies include a "shield" chest, widely spaced nipples, short and webbed neck, cubitus valgus, and Madelung deformity. Neonates may have congenital lymphedema of the hands and feet, webbed neck, nail dysplasia, narrow and high-arched palate, and short fourth metacarpals and/or metatarsals. Cardiovascular disease, Ovarian failure, renal abnormalities and hypertension, increased risk for gonadoblastoma (as in 45,X/46,XY mosaicism), Autoimmune disorders (Thyroid disease, Celiac, IBD), are other disorders that need periodical Screening. Intelligence is usually within the normal range; some may have specific neurocognitive deficits. Birth length is approximately -1 SD. Linear growth remains slow, further compounded by delayed puberty, with lack of a pubertal growth spurt. Unfortunately, many girls, in spite of their short stature, are not diagnosed until approximately age 8-10 years (or later), years after the patient falls below the fifth percentile for height, which is too late to derive full benefit from treatment with growth hormone. GH, Estradiol therapy, oxandrolone & Adjunctive progestins with estradiol, are among most drugs use for short stature or ovarian failure treatment in these patients.





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#### normal puberty development

## Fatemeh Sayarifard <sup>1</sup> © P

**Abstract**: Puberty refers to the physical changes that occur during adolescence. Puberty consists of a series of predictable events in physical, cognitive and psychosocial maturation, with some variation in timing of onset, sequence, and tempo. In girls, the first signs of puberty onset are with a wide range from approximately 8 to 13 years in healthy girls. Pubertal onset in girls has been trending earlier in most countries. This rate varies by weight status and race/ethnic group. In boys, the first signs of pubertal onset are with a range from approximately 9 to 14 years, with some variation among race/ethnic groups. Precocious puberty is defined as pubertal onset at an age 2 standard deviations (SD) below the mean age of onset of puberty. Similarly, delayed puberty is defined as lack of pubertal onset by an age2SD above the mean age of onset of puberty. Sexual maturity ratings based on Tanner stages for breast in girls, male genitalia and pubic hair, consist of five categories. (stage1: prepubertal, stage 5: adult form, stage 2,3,4 between them). The growth spurt occurs approximately two years earlier in girls than in boys. Puberty is associated with significant changes in body weight and alterations in body composition, especially in lean body mass and the proportion of body fat, with different patterns in girls as compared with boys. Conclusion: The most visible changes during puberty are growth in stature and development of secondary sex characteristics. Equally profound are changes in body composition; achievement of fertility; and changes in most body systems such as bone (with increased growth and mineralization), brain (with ongoing development), and the cardiovascular system (with greater aerobic power reserve, electrocardiographic changes, and blood pressure changes). While the hormonal changes that drive pubertal development are well described. The physiologic mechanisms that determine pubertal timing remain poorly understood.





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## **Precocious puberty**

حسين شعباني ميرزائي  $^1$  ©  $^0$ 

**Abstract**: Definition of precocious puberty is appearance of physical and hormonal signs of pubertal development at an unusually early age. Precocious puberty can have adverse consequences which include the following: reduced adult height potential, effects on social behavior and psychological development, and possibility of causing some lifelong health risks. Puberty is described as precocious in girls that onset of puberty happening before age 8 years. In boys, when precocious puberty is denominated that pubertal development occurred before age 9 years. Premature thelarche is common, benign, normal variant conditions that can resemble true precocious puberty but that progress slowly or not at all. Premature thelarche refers to the isolated appearance of breast development, usually in girls younger than 3 years. If the history and physical examination suggest that a child has sign of pubertal maturation, the clinician must determine whether this is a precocious puberty or not. In case of precocious puberty, it is necessary to differentiate between central precocious puberty (CPP) and peripheral precocious puberty (PPP). In CPP, which is gonadotropin-dependent, early maturation of the entire hypothalamic-pituitary-gonadal (HPG) axis occurs, with the full spectrum of physical and hormonal changes of puberty. Peripheral precocious puberty is much less common and refers to conditions in which increased production of sex steroids is gonadotropin-independent. Correct diagnosis of the etiology of sexual precocity is critical because the evaluation and treatment of patients with peripheral precocious puberty are quite different from those of patients with CPP. In addition to history and physical examination, paraclinical evaluations are also used for diagnosis. Measurement of serum sex hormones is useful in cases with suspected precocious puberty. Imaging studies can be helpful in precocious puberty evaluation. Based on the etiology and type of precocious puberty, medical or surgical treatments are used.





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## Type 2 diabetes in children



**Abstract**: Type 2 diabetes is a progressive disorder due to a deficit in both insulin secretion and insulin action, with obesity being the primary cause in children. Data indicates that the incidence of type 2 diabetes is rising. Often asymptomatic and diagnosed by screening in a high-risk individual (e.g., family history, obesity, acanthosis nigricans) or incidentally (e.g., glycosuria found during a school or sports exam). Obesity and excess adipose tissue especially when centrally distributed are the most important risk factors. Family history of gestational diabetes is also strong risk factor. Diagnosis is based on fasting plasma glucose of more equal 126 mg/dL, random plasma glucose of more equal 200 mg/dL with symptoms of hyperglycemia (e.g., polyuria or polydipsia) or hyperglycemic crisis, a plasma glucose level of more equal 200 mg/dL 2 hours after 75 g oral glucose, or hemoglobin A1C of more equal 6.5%. In the absence of unequivocal hyperglycemia, the test must be repeated to substantiate the diagnosis. The definitions of prediabetes: A fasting plasma glucose level of 100 to 125mg/dl, an HbA1c level of 5.7% to 6.4% or a 2 hour post load glucose level of 140 to 199 mg/dl are consistent with prediabetes. Obesity, leading to insulin resistance, is the primary cause in children. The development of insulin resistance and glucose intolerance can be prevented or delayed by lifestyle modifications that correct obesity in children. Goals of treatment are to promote weight loss and exercise capacity, decrease acanthosis nigricans, normalize glycaemia and hemoglobin A1C (goal is less than 7%), and prevent long-term complications and comorbidities (e.g., retinopathy, hypertension, and dyslipidemia). Initial treatment includes lifestyle modifications, metformin, and insulin. Liraglutide and exenatide are approved in some countries as an additional non-insulin treatment option for children ages more equal 10 years.





#### Chronic Diarrhea



Abstract: Chronic Diarrhea Mehri Najafi MD Professor in pediatric Pediatric gastroenterologist Diarrhea is the excessive loss of fluids and electrolytes in the stool. When the duration of diarrhea lasts more than 14 days it is defined as chronic and persistent diarrhea. The worldwide prevalence of chronic diarrhea ranges from 3 to 20%. Though incidence of diarrhea has not decreased, the mortality is decreasing due to development of rotavirus vaccine , nutritional states, and medical substance. The causes of chronic diarrhea are divided into infectious and noninfectious etiologies. Others classify chronic diarrhea according to pathophysiology, and the presence or absence of failure to thrive. Causes of chronic diarrhea according to presence or absence of FFT Without FTT With FTT Chronic nonspecific diarrhea Allergic enteropathy Infectious colitis Intractable diarrhea of infancy Lactose malabsorption Celiac disease Small bowel bacterial overgrowth Inflammatory bowel disease Irritable bowel syndrome Immunodeficiency state Cystic fibrosis Autoimmune enteropathy The most frequent cause of chronic diarrhea is enteric infection. However incidence of autoimmune enteropathy and IBD is increasing. Chronic nonspecific diarrhea is the most common cause of chronic diarrhea during 1 to 3 years of age. These patients pass stools only during waking periods and keep healthy state without weight loss and nutritional deficits. The causes are increased intestinal motility and osmotic effects in GI tract, due to solutes that are high in carbohydrate content. Chronic diarrhea in early infant periods has the characteristics of high mortality and morbidity. They divided into two groups, 1) without villous atrophy, and with villous atrophy. Microvillous inclusion disease, tufting enteropathy and autoimmune enteropathy have with villous atrophy. Congenital transport defects (congenital chloride diarrhea) congenital glucose galactose malabsorption villous are intact.





## Clinical symptoms of chronic diarrhea

# Mandana Rafeey <sup>1</sup> © P

**Abstract**: Objective definitions of diarrhea are based on stool weight or volume. Thus, diarrhea can be defined by one or both of the following measures: Stool weight: Daily stool weight greater than 250 g in children that weigh more than 10 kg is considered diarrhea. In young children less than 10 kg stool over 20 g/kg/day is abnormal. Stool consistency: The Bristol stool chart helps define stool consistencies. Diarrheal stools typically correspond to types 6 and 7 on the Bristol stool chart. However, if the stool volume is low, isolated loose stools are not a significant predictor of underlying disease in the absence of weight loss, dehydration, or significant changes in biochemical/nutritional indicators. Chronic diarrhea – Chronic diarrhea is generally defined as diarrhea lasting more than four weeks. This timeline is selected to distinguish between the acute diarrheal episode, which tends to be self-limited, and more prolonged diarrheal disease that warrants further evaluation and possibly intervention. Persistent diarrhea – Persistent diarrhea is sometimes defined as diarrhea lasting longer than two weeks, but, other times, this term is used interchangeably with chronic diarrhea. Warning signs: The first step is to identify any warning signs or symptoms suggesting a rapidly progressive or severe systemic disorder. Children with warning signs may require hospital admission and an expedited evaluation. Key warning signs are: Fever, Gross blood in stool, Growth faltering or weight loss, Nausea, vomiting Severe abdominal distension or tenderness. Hepatosplenomegaly or mass Key examination findings for a child age up to 6 months with chronic diarrhea, Physical examination findings Implications: Aphthous stomatitis IBD and celiac disease Stomatitis/glossitis/angular cheilitis B-complex vitamin deficiency secondary to malabsorption Diaper/perianal rash Acidic stools – Carbohydrate malabsorption Acrodermatitis enteropathica – Zinc deficiency Cutaneous rashes Dermatitis herpetiformis – Celiac disease Erythema nodosum – IBD Pyoderma gangrenosum – IBD Loss of subcutaneous fat





## Complementary nutrition

<sup>1</sup> © ® فريبا صيقلى

هدف و ضرورت تغذیه تکمیلی شیر خوار: تامین ماکرو و میکرو نوترینت های لازم برای رشد شیر خوار :پایان شش ماهگی، آمادگی برای مصرف غذاهای خانواده پس از پایان یک سالگی و قطع شیر دهی پس از پایان دو سالگی، آمادگی مصرف غذای نیمه جامد و سپس جامد, شکل گیری ذائقه و تحمل طعم ها و مزه های مختلف، تقویت و شکل گیری مهارت جویدن و بلع غذای جامد. یکی از وظایف مهم سیستم های بهداشتی تربیت مشاورین تغذیه در مراکز مراجعه مادران به علم و مهارت کافی در این زمینه هست. اصول کلی تغذیه تکمیلی تغذیه: مصرف غذا در سال اول عمر بعد از مصرف شیر مادر یا شیر خشک انجام شود. ۳ تا ۵ روز فاصله بین غذاهای جدید اعمال شود تا آلرژی یا عمر بعد متی تا ۱۰ بار. شروع هر ماده غذایی با مقدار خیلی کم و سپس افزایش یابد. غلظت و قوام غذای تکمیلی شروع شود، حتی تا ۱۰ بار. شروع هر ماده غذایی با مقدار خیلی کم و سپس افزایش یابد. غلظت و قوام غذای تکمیلی متناسب با سن شیر خوار از کمی غلیظ تر از شیر و به مرور غلظت افزایش داده شود،افزودن نمک یا شکر در ابتدای متناسب با سن شیر خوار از کمی غلیظ تر از شیر و به مرور غلظت افزایش داده شود،افزودن نمک یا شکر در ابتدای مختلف دارند مثلا شروع لبنیات. برخی دستورات غذایی مربوط به منطقه جغرافیایی و یا سلیقه خانواده هست. مثلا عدم مصرف برنج در شروع تغذیه در مناطقی که برنج آلوده به سموم آرسنیک هست و یا سیب زمینی و هویج که آلودگی با مصرف برنج در شروع تغذیه در مناطقی که برنج آلوده به سموم آرسنیک هست و یا سیب زمینی و هویج که آلودگی با ماهه اقدام می شود





## Diagnosis of gastroesophageal reflux disease in children

## Shohreh Maleknejad MD 1 © P

**Abstract**: Gastroesophageal reflux related symptoms are a major cause of parental concern and referrals at all ages. When children present with symptoms suggesting GERD, further work-up is appropriate. Selection of tests depends upon the clinical scenario. Contrast radiography may be useful to exclude anatomic abnormalities in selected patients, such as those with dysphagia or odynophagia, young infants with intractable reflux, or those with suspected obstruction. Ultrasonography is not indicated for GERD diagnosis as the results are clearly investigatordependent. The specificity of ultrasound in the 15 minutes postprandial is only 11% in comparison to pH-metry. Esophageal Manometry has poor specificity and sensitivity in the diagnosis of GERD. It is useful to rule out esophageal motility disorders during pre-operative evaluation of children undergoing fundoplication. Scintigraphy is a radionuclide based study for the diagnosis of pulmonary aspiration in GERD. It has low sensitivity, lacks standardized technique and there are no accepted normal values. Endoscopy of the upper GI tract is useful to evaluate the mucosa in the presence of alarm symptoms or signs, such as hematemesis, dysphagia, or failure to thrive or anemia; to detect complications of GERD, such as erosive esophagitis, strictures, and Barrett's esophagus; or to diagnose conditions that might mimic GERD, such as eosinophilic esophagitis. Biomarkers, such as salivary pepsin, have not been shown to be useful to diagnose GERD. Empirical therapy has no benefit in infants, while a 4 week trial can be given in older children with symptoms suggestive of esophagitis. Esophageal pH and impedance monitoring studies may be useful for patients with atypical symptoms, atypical asthma, or extra-esophageal symptoms, but generally not for a child with typical heartburn. CONCLUSION: A careful history and clinical examination are adequate to make a diagnosis in most patients, but judicious investigations are necessary in a few.





#### **GERD**

# دكتر فرزانه معتمد $^1$ © $^{ ext{P}}$

دانشگاه علوم پزشکی تهران ۱

Abstract: Gastroesophageal reflux (GER): • Passage of gastro/duodental contents into the esophagus • Physiologic: common in infants, postprandially in all ages Gastroesophageal reflux (GERD): • Reflux causing complications, sometimes with objective damage (esophagitis, stricture, and barrettes) or systemic, eg, failure to thrive • Reflux causing subjective troublesome symptoms, eg, heartburn or regurgitation or vomiting above some threshold. Regurgitation/"spitting up'/spilling": • Reflux into oropharynx or effected from mouth, usually effortless/no forceful but may be quite forceful, especially in infants. • Sometimes termed 'vomiting' Vomiting: • Forceful ejection of gastro/duodenal contends form mouth Red flags of GER: • FTT • Nausea • Vomiting • Hematemesis • Difficulty in swallowing • Bilious vomiting • Fever • Organomegaly • Lethargy • Apnea If there is alarm signals or red flags, the GER is secondary. Then metabolic disorders in young children, Peptic diseases, gastrointestinal obstruction, Pancreatic, Hepatic, Biliary diseases, should be ruled out. Protection of the esophagus from GERD: • Lower esophageal sphincter • Esophageal capacitance and clearance • Mucosal mucus and bicarbonate secretion • Swallowed saliva buffering residual acid Protection of the airway from GERD: • Upper esophageal sphincter • Esophageal –glottal closure reflux • Reflux apnea • Pharyngeal clearance • Cough • Ciliary airway clearance Physiologic reflux does not need any evaluation and history and physically exam is enough for diagnosis. For pathologic or secondary reflux we should do more evaluation. Evaluation: • Empiric treatment • Endoscopy and histology • Esophageal pH monitoring or impedance monitoring • Barium contrast radiography • Bronchoalveolar lavage • Nuclear scintigraphy Atypical symptoms: • Nonallergic asthma • Chronic cough • Hoarseness • Pharyngitis • Chest pain (mimics angina) • PND • May be only symptoms - 'omeprazol test' Treatment: • Lifestyle change • Acid suppressing medications • PPI: there is acid rebound after abrupt stop • H2RA: short term

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#### **GERD Management**

يريسا رحماني  $^1$   $^{\odot}$   $^{\odot}$ 

Abstract: GERD MANAGEMENT 1-Lifestyle changes: • Head of bed elevation and left lateral positioning, Avoid foods that tend to induce reflux by reducing lower esophageal sphincter pressure, including chocolate, peppermint, and caffeinated beverages. • Avoid acidic foods, including colas, orange juice, and tomato sauce. Although their contribution to gastric acidity is probably minimal. • Avoid high-fat foods in selected patients. This strategy is recommended in adults with GERD because fatty foods tend to slow gastric emptying and thereby promote reflux. Decisions about whether to restrict dietary fat depend on the infant or child's overall nutritional status and the clinical response • Weight loss or weight management for individuals who are overweight • Others incluing (Positioning, Salivation, Avoid alcohol and tobacco) 2-Pharmacotherapy • Proton pump inhibitors (PPIs) (for Esophagitis or marked GERD symptoms & Asthma and other GERD extra esophageal manifestations) • Histamine type 2 receptor antagonists (H2RAs) (For patients with mild or intermittent symptoms of GERD • Antacids(for short-term relief of heartburn) • Surface agents • Prokinetics 3-Surgery ( Gastrostomy placement ,Fundoplication, endoscopic radiofrequency ablation of the lower esophageal sphincter, endoscopic full-thickness fundoplication, and total esophagogastric disconnect ) MD.PARISA.RAHMANI

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## **Neonatal Hepatitis (cholestasis)**

<sup>1</sup> © P دکتر فاطمه فرهمند

دانشگاه علوم پزشکی تهران <sup>۱</sup>

**Abstract**: The term neonatal hepatitis originated in the 1950s when few causes of neonatal liver disease were identified, and pathologists recognized a characteristic histologic appearance of the neonatal liver in response to injury. The term has since been used to refer to virtually all forms of liver dysfunction in the neonate that present clinically as jaundice due to conjugated hyperbilirubinemia within the first 3 months of life. The neonatal liver thus results in further impairment of the excretory machinery, leading to clinically significant jaundice. For practical purposes, neonatal cholestasis is defined as a conjugated hyperbilirubinemia. Conjugated bilirubin fraction greater than 20% of the total serum bilirubin level or serum conjugated bilirubin greater than 1 mg/dL. A term proposed to circumvent these imprecisions is neonatal hepatitis syndrome, which emphasizes the uniformity of the clinical phenotype caused by the conglomerate of infectious, genetic, toxic, and metabolic causative and anatomic abnormalities including extrahepatic biliary atresia disease processes leading to impaired excretory function and bile secretion ,leading to the common phenotype of pathologic cholestasis in the neonate and infant. The North American Society for Pediatric Gastroenterology, Hepatology and Nutrition Cholestasis Guidelines recommends that all infants who are jaundiced at 2 weeks of age (or 3 weeks if breast-fed and with normal history and no pale stools or dark urine) be screened for cholestasis with measurement of fractionated serum bilirubin. Disorders associated with cholestasis in the neonate are diverse, although the clinical presentation is similar, early recognition of cholestasis in the infant and prompt identification of the treatable disorders such as extrahepatic biliary atresia, sepsis, endocrinopathies and specific metabolic disorders (such as galactosemia, tyrosinemia type I), allow initiation of appropriate treatment to prevent progression of liver damage and, if possible, reverse damage that has already occurred. Key word:cholestasis, conjugated hyperbilirubinemia,neonate





# pathogenesis and etiology of pediatric cholestatic liver disease

<sup>1</sup> © هناز صادقیان

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**Abstract**: Pathogenesis of cholestatic liver diseases Cholestatic liver diseases begin to develop after an impairment of bile flow start to affect the biliary tree and subsequently hepatocytes. Several factors cause bile flow disturbances. Antigenic stimuli, exotoxins, endotoxins, xenobiotics, Early cholangiocyte response may allow resolution of injury, however, sustained pro-inflammatory signaling associated with deregulation of genetic and/or epigenetic regulatory mechanisms could condition late dysfunctional permanent state. There are 3 important aspects in cholestasis pathogenesis. Ductular reaction is the first step that size, metabolic rate as well as proliferative and plasticity capabilities varies due to triggering factors. hepatocytes and cholangiocytes have a common stem cell precursor, and trans differentiation may occur in massive parenchymal loss. The second step is Bile acid toxicity and mitochondrial dysfunction which Besides its functional role of converting lipid bilayers into mixed micelles, BA are endogenous ligands that activate a network of receptors, that provide an inflammatory signaling in liver response to injuries. Intrahepatic BA can further be processed by hydroxylation, glucuronidation or sulfation, and excreted back into sinusoidal and systemic circulation. Critical steps in the enterohepatic circulation are regulated by the BA receptor, which limits BA uptake and synthesis by enhancing biliary and basolateral BA export. The third fundamental aspect of the core framework is the influence of immunogenetics and epigenetics on immunoinflammatory response. Patients with CLD exhibit a variety of genetic alterations that account for the different elements of each CLD. However, some of those genes may be directly implicated in the progression rate of the cholestatic phenotype. To complete the core framework of CLD, dysfunctional matrix rearrangements and fibrogenesis are the fourth concept. Fibrogenesis is a dynamic process that appears immunoinflammatory mechanisms, secretion of tissue metalloproteinases, cytokine networks and derangements of mesenchymal cells infiltration with ultimate loss of tissue maintenance homeostasis.

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## Pathophysiology of gastroesophageal reflux

كامبيز افتخارى  $^1$   $^{\odot}$   $^{\odot}$ 

دانشگاه علوم پزشکی تهران <sup>۱</sup>

**Abstract**: Pathophysiology of gastroesophageal reflux Gastroesophageal reflux (GER) refers to the involuntary passage of gastric contents into the esophagus. Most episodes of GER are brief, asymptomatic, and limited to the distal part of the esophagus. Reflux disease (GERD) occurs when the reflux episodes are associated with symptoms or complications. The lower esophageal sphincter (LES) acts as a valve and it is considered an anti-reflux barrier. The frequency of reflux episodes is increased by insufficient LES tone, abnormal frequency of LES relaxations, and by hiatal herniation. Transient lower esophageal sphincter relaxation (TLESR) is the most important pathophysiologic. Gastric distention is the main trigger for TLESR. Other factors such as increased movement, straining, obesity, hyperosmolar and large-volume meals, gastroparesis, a large hiatal hernia, and increased respiratory effort (coughing, wheezing) can also have similar effects. Three major lines of defense to minimize esophageal damage secondary to reflux: first, the anti-reflux barrier is the result of the LES, the diaphragm and its angle of His. The second is the peristalsis and clearance of the esophagus and the third is the resistance of the tissue and mucosa of the esophagus. The duration of reflux episodes is increased by lack of swallowing (during sleep) and by defective esophageal peristalsis. Healthy and sick individuals do not differ in the presence or absence of GER but rather in the frequency, duration, and intensity of GER and in its association with symptoms or complications. Therefore, Factors determining the esophageal manifestations of reflux include the duration of esophageal exposure (a product of the frequency and duration of reflux episodes), the causticity of the refluxate, and the susceptibility of the esophagus to damage.





## Sighn and symtoms of Cholestasis

#### Abolfazl Iranikhah <sup>1</sup> © P

**Abstract**: Clinically, cholestasis in the infant may present as jaundice, pruritus, fat-soluble vitamin deficiency, or may evolve during or following acute liver failure. Functional or anatomic biliary obstruction is often heralded by the presence of acholic stools. Although cholestasis is frequently the primary presenting symptom of neonatal hepatobiliary disease, it also commonly represents the final common pathway of any disease that affects the neonatal liver. As such, cholestasis is often classified by origin and is designated as either (1) biliary, referring to structural abnormalities and obstruction of extrahepatic or intrahepatic bile ducts; or (2) hepatocellular, resulting from impairment in bile transport, genetic or metabolic abnormalities, and infection. Biliary atresia (BA) presents most commonly with cholestasis between 2 and 5 weeks of life. Acholic stools may be present and indicate biliary obstruction; however, onset commonly follows the onset of jaundice. Treatable conditions that can present with cholestatic iaundice include bacterial sepsis, galactosemia, tyrosinemia, panhypopituitarism, bile acid synthetic defects, or obstructive gallstones. These infants often appear acutely ill and early diagnosis will enable timely initiation of directed treatment. Conversely, infants with BA usually appear otherwise healthy and grow normally which may deceive the parent or physician into believing that the jaundice is physiologic or caused by breast-feeding. The clinician performing a physical examination should not only focus on the abdomen but should also consider extra hepatic signs, such as: dysmorphic features, poor growth, dermatologic, neurologic, or pulmonary symptoms. Palpation of the abdomen may reveal firm hepatomegaly suspicious for the diagnosis of BA, often with a prominent middle or left lobe. Splenomegaly in BA appears after the newborn period, and if present at a young age of 2 to 4 weeks should point toward other diseases such as storage or hematologic disorders. Cardiac examination is the key, as discovery of a murmur may suggest





## Steps to support breastfeeding and the role of pediatrician

دکتر محمود راوری  $^1$  ©  $^{ ext{P}}$ 

دانشکده علوم پزشکی ساوه - عضو کمیته کشوری شیرمادر - عضو هیئت مدیره انجمن علمی ترویج تغذیه با شیرمادر ۱

**Abstract**: Breastfeeding and human milk are the normative standards for infant feeding and nutrition. The short and long-term medical and neurodevelopmental advantages of breastfeeding make breastfeeding, or the provision of human milk, a public health imperative. The American Academy of Pediatrics (AAP) recommends exclusive breastfeeding for approximately 6 months after birth. Furthermore, the AAP supports continued breastfeeding, along with appropriate complementary foods introduced at about 6 months, as long as mutually desired by mother and child for 2 years or beyond. Support of breastfeeding Begins during Pregnancy that is helpful to have the obstetric health care provider acknowledge support for breastfeeding early in the pregnancy and Breastfeeding Promotion in the Prenatal Setting. Breastfeeding National, State, Local Support Infrastructure and Hospital support is very important. All hospitals are encouraged to adopt the Ten Steps for Successful Breastfeeding recommended by the WHO and endorsed by the AAP. Implementation of at least 5 of the Baby-Friendly hospital practices, including, including Breastfeeding in the first hour after birth, Exclusive breastfeeding, Rooming in, Breastfeeding on demand, Avoidance of pacifiers, and Information on breastfeeding support after discharge, Enabled women to be more successful at meeting their prenatal desire for exclusive breastfeeding. Formal hospital staff training should focus not only on updating knowledge and techniques for breastfeeding support but also should acknowledge the need to change attitudes about the equivalency of breastfeeding and commercial infant formula feeding. Pediatricians can play an important role in leading and advocating for the societal changes that permit continued exclusive and direct breastfeeding. Pediatricians play a critical role in hospitals, their practices, and communities as advocates of breastfeeding and, thus, need to be trained about the benefits of breastfeeding for mothers and children and in managing breastfeeding.





# SYMPTOMS and SIGNS ASSOCIATED WITH GEREFLUX DISEASE:



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**Abstract**: The presenting symptoms and sigs of GERD differ according to age. Infantile reflux manifests more often with regurgitation (especially postprandially), signs of esophagitis (irritability, arching, choking, gagging, feeding aversion), and resulting failure to thrive; symptoms resolve spontaneously in the majority of infants by 12-24 months. Older children can have regurgitation during the preschool years; this complaint diminishes somewhat as children age, and complaints of abdominal and chest pain supervene in later childhood and adolescence. Occasional children present with food refusal or neck contortions (arching, turning of head) designated Sandifer syndrome. The respiratory presentations are also age dependent: in infants may manifest as obstructive apnea or as stridor or lower airway disease. Otitis media, sinusitis, lymphoid hyperplasia, hoarseness, vocal cord nodules, and laryngeal edema have all been associated with GERD. Airway manifestations in older children are more commonly related to asthma or to otolaryngologic disease such as laryngitis or sinusitis. Atypical symptoms such as epigastric pain, nausea, flatulence, hiccups, chronic cough, asthma, chest pain, and hoarseness account for 30% to 60% of presentations of GERD. Possible associations exist between GERD and asthma, pneumonia, bronchiectasis, acute lifethreatening event, laryngotracheitis, sinusitis, and dental erosion. Clinicians need to be aware that not all regurgitation and vomiting in infants and young children is related to GER/GERD. Bilious vomiting, gastrointestinal bleeding, consistently forceful vomiting, weight loss or failure to thrive, diarrhea, constipation, fever, lethargy, hepatosplenomegaly, abdominal tenderness, and/or distension should raise the possibility of an alternate diagnosis. The rapidly increasing prevalence of obesity is causing a rising prevalence of GERD. The risk of GERD symptoms is associated with the increase in body mass index and waist circumference, even in normal-weight children.





#### treatment of chronic diarrhea

دكتر حامد شفق  $^1$  ©  $^2$ 

دانشگاه علوم پزشکی آزاد اسلامی تهران ۱

Abstract: TREATMENT OF CHRONIC DIARRHEA HAMED SHAFAGH, MD. **PEDIATRIC** GASTROENTEROLOGIST ASSOCIATE PROFESSOR. **TEHRAN** MEDICAL UNIT, ISLAMIC AZAD UNIVERSITY Treatment includes general supportive measures, nutritional rehabilitation, elimination diet, and drugs. Because death in most instances is caused by dehydration, replacement of fluid and electrolyte losses is the most important early intervention. Rehydration is best performed with ORS. Super ORS provide advantages over the pure glucose and electrolyte conventional WHO ORS. Nutritional rehabilitation is essential and in moderate to severe malnutrition, caloric intake may be progressively increased to 50% or more above the recommended dietary allowances for age and sex. In children with steatorrhea MCT should be the main source of lipids. A lactose free diet should be started in all children with chronic diarrhea. Semi elemental or elemental diets are used in food intolerance. The sequence of elimination should be graded from less to more restricted diets, that is, cow's milk protein hydrolysate to amino acid based formula, depending on the child's situation. In severely compromised infants, it may be convenient to start with amino acids based feeding. The route of feeding may be enteral or parenteral based on clinical situations. Micronutrient and vitamin supplementation are part of nutritional rehabilitation, such as zinc. Drug therapy includes anti infectious drugs, immunosuppression and drugs that may be inhibit fluid loss and promote enterocyte growth. Co-trimoxazole, metronidazole or albendazole, and nitazoxide have a broad pattern of target agents, including parasites. In rotavirus, induced severe and protracted diarrhea, oral administration of IVIG should be considered. Immunomodulator agents, such as azathioprine, cyclosporine and tacrolimus, have been used in severe protracted diarrhea of immune origin. Octerotide may be effect in severe secretory diarrhea. Other drugs are loperamide, chlorpromazine, zinc, growth hormone promotes, butyrate and probiotics. Other disease such as IBD, CF, liver disease,... have specific treatment.

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# Clinical guidelines for the diagnostic and therapeutic management of monkey-pox in children

# Mahya sadat Mohammadi <sup>1</sup> © P

**Abstract**: Monkey pox disease is a type of viral disease that is caused by the DNA virus belonging to the orthopoxvirus genus of the Poxviridae family. A case defined as a suspected or probable case and monkeypox virus by diagnostic methods, isolation of monkeypox virus in culture or demonstration of monkeypox virus DNA by polymerase chain reaction (PCR) test of a clinical specimen or morphological demonstration of the virus with orthopox The virus is isolated by electron microscopy in the absence of exposure to other orthopox virus or showing the presence of orthopox virus in the tissue using immunohistochemical test methods in the absence of exposure to other orthopox virus, has a definitive diagnosis of this disease. Transmission of monkeypox virus (Monkeypox virus; MPXV) occurs when a person comes into contact with the virus through an animal, human or material infected with the virus. The virus enters the body through broken skin (even if it is not visible), the respiratory tract, or mucous membranes (eyes, nose, or mouth). Symptoms of this disease include fever, severe headache, lymphadenopathy, myalgia. Skin lesions usually begin within 1 to 3 days after the onset of fever and include macules, papules, vesicles, and pustules. Complications of the disease include bacterial skin infection, permanent skin scarring, hyperpigmentation or hypopigmentation, permanent corneal scarring, pneumonia, dehydration, sepsis, encephalitis, eye complications. Many people infected with monkeypox virus have a mild and self-limiting disease course in the absence of specific treatment. Recovery usually occurs in 2-4 weeks. The main prevention strategy is to increase awareness of risk factors and educate people to reduce exposure to the virus. Some countries have policies to provide vaccines to people who may be at risk, such as laboratory personnel, rapid response teams, and health workers.







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#### Covid-19 vaccine in children

# Shirin Sayyahfar <sup>1</sup> © P

Vicalization Communication Com

**Abstract**: Following the covid-19 pandemic and the production of the vaccines against this virus, now eligible children must also receive this vaccine like adults. The CDC approves the Pfizer and Moderna vaccines for use in children starting at 6 months of age. In Iran, Sinopharm and PastoCoVac vaccines are intended for children from the age of 5 years and above. Studies show that the administration of the covid vaccine in children reduces the rate of hospitalization due to this disease and can provide protection against MIS-C. The covid vaccine can be prescribed with all other vaccines at the same time or at any interval. It is not recommended to perform serological tests to decide to administer the vaccine or the booster dose or to check the response of the immune system after the administration of the vaccine. Children who have already been infected with the covid 19 disease should also receive the vaccine like non infected ones. Since the risk of re-infection is low up to 3 months after infection, the administration of the vaccine can be postponed until 3 months after infection. In order to administer the covid vaccine to a child with history of MIS-C due to covid 19 disease, an individual decision should be made. Considering that the exposure to this virus has led to an immune dysregulation in the individual, it is possible that the administration of the vaccine will also cause such a response with the same mechanism. The contraindication of this vaccine is a severe allergic reaction such as anaphylaxis to the previous covid-19 vaccine dose or to a component of the vaccine. It should be noted that usually the development of hives after 4 hours from the time of injection is usually not related to the vaccine.





oral

#### **Fever**

#### Iran Malekzadeh <sup>1</sup> © P

**Abstract**: Fever is one the most complaint in children (15–25% of emergency ward referrals). It is defined as rectal temperature more than 38 centigrade degree. At the first step we have to decide to admit a patient or treat him as an outpatient, these are some criteria for admission: illness, instability, prolonged fever, present of risk factor, etc. Pediatricians categorize children with fever into 3 groups by age: 1. younger than 90 day, 2. 3 months to 3 years, 3. older than 3 years old. In a simple vision all febrile neonates (younger than 30 days) must be admitted, on the other hand, it is very easy to detect the source of fever in a child older than 3 years, whom we have to exam carefully. In the middle group (3 mo.- 3 Y) pediatricians refer to vaccination history and risk factors for urinary tract infections. Some paraclinical evaluations such as lab data (CBC, B/C, ESR, CRP, U/A) and chest x-ray may help diagnose the reason for the fever. Considering the source of the fever, the patient's age and the duration of the disease, treatment varies from antipyretic to intravenous infusion of wide-spectrum antibiotics.

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#### Human papillomaviruses vaccines

Dr Zahra Movahedi <sup>1</sup> © ®

**Abstract**: Three different vaccines, which vary in the number of HPV types they contain and target, have been clinically developed, •Quadrivalent vaccine (Gardasil) targets HPV types 6, 11, 16, and 18. • Nine-valent vaccine (Gardasil 9) targets the same HPV types as the quadrivalent vaccine (6, 11, 16, and 18) as well as types 31, 33, 45, 52, and 58. •Bivalent vaccine (Cervarix) targets HPV types 16 and 18. These are all prophylactic vaccines, designed to prevent initial HPV infection and subsequent HPV-associated lesions. HPV types 16 and 18 which are targeted by all three HPV vaccines cause approximately 70% of all cervical cancers worldwide, 90% of anal cancers and substantial proportion of oropharyngeal and penile cancers and HPV types 31, 33, 45, 52, and 58, which are additionally targeted by the 9-valent vaccine, cause an additional 20 percent. Vaccination with the quadrivalent or 9-valent HPV vaccine also protects against anogenital warts (90 percent of which are caused by HPV types 6 and 11). Vaccinating both males and females is more beneficial in reducing HPV infection and disease than by vaccinating only females • Routine HPV vaccination is recommended at 11 to 12 years. It can be administered starting at 9 years of age. •For adolescents and adults aged 13 to 26 years who have not been previously vaccinated or who have not completed the vaccine series, catch-up vaccination is recommended. •For adults 27 years and older, catch-up vaccination is not routinely recommended. For people initiating vaccination before their 15th birthday the recommended schedule is 2 doses at 0, and 6 to 12 months after the first dose. For people initiating vaccination on or after their 15th birthday, the recommended schedule is 3 doses at 0, 1 to 2(typically 2), and 6 months after first dose





oral

#### Influenza vaccine

#### Mahmoud Khodabandeh <sup>1</sup> © P

Medical Center, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran.

**Abstract**: The word influenza was first used in English to refer to the disease we know today in 1703. The most famous and lethal outbreak was the 1918 flu pandemic (Spanish flu pandemic) (type A influenza, H1N1 subtype), which lasted from 1918 to 1919. The WHO each year releases a recommended formula for each region of the world based on the data it has collected on the prevalence of particular strain. The current influanza vaccines are contained the most prevalent types of inf. A and inf. B In the last two decades, four major and at least eight minor mismatches between vaccine and circulating B viruses have occurred in the northern hemisphere, thus impairing the performances of TIVs. In Europe, a B-mismatch between vaccine and circulating strains occurred in 5 of 10 seasons between 2001 and 2011. In 2012 the WHO recommended the production of quadrivalent influenza vaccines (QIVs) for seasonal immunization For protection against the influenza B lineage not present in the trivalent vaccine. Routine annual influenza vaccination is recommended for all persons aged ≥6 months who do not have contraindications. When vaccine supply is limited emphasis should be placed on vaccination of high-risk groups and their contacts/caregivers. Contraindications infulanza vaccine: 1- Children younger than 6 months 2- Individuals with severe, life-threatening allergies to flu vaccine or any ingredient(s) in the vaccine 3- Allergy to eggs or any of the ingredients in the vaccine 4- Guillain-Barré syndrome (GBS) during 6week after previous dose All person require 1 dose of influenza vaccin annually. Children who needs 2 doses: at least 4 weeks apart. Children aged 6 months through 8 years who have never been vaccinated against influenza or for whom vaccination history is unknown. Children aged 6 months through 8 years who have not received at least 2 doses of seasonal influenza vaccine





oral

## **Principles of Antimicrobial Prophylaxis**

خدیجه دانشجو $^1$  ©  $^{ ext{P}}$ 

هیئت علمی کودکان علوم پزشکی تهران ۱

**Abstract**: Principles of Antimicrobial Prophylaxis Antimicrobial prophylaxis is defined as the use of antimicrobial drugs in the absence of suspected or documented infection to prevent development of infection or disease and is a common practice in pediatrics. Although the efficacy of antimicrobial prophylaxis has been demonstrated for some conditions, this is not the case for many more conditions for which it is nevertheless used. Concerns about the emergence of resistant bacterial pathogens has led to a reexamination of the role of antimicrobial prophylaxis, especially for conditions for which prolonged administration is required, such as prevention of recurrent otitis media (OM) and urinary tract infection (UTI). Effective chemoprophylaxis should be directed at pathogens common in the infection-prone body sites. When using prophylactic antimicrobial therapy, the risk of the emergence of antimicrobial-resistant organisms and the possibility of an adverse event from the drug must be weighed against potential benefits. Ideally, prophylactic agents should have a narrow spectrum of activity and should be used for a brief period of time. Infection-Prone Body Sites Antibiotic prophylaxis in vulnerable body sites is most successful if: (1) the period of risk is defined and brief; (2) the expected pathogens have predictable antimicrobial susceptibility; and (3) the site is accessible to adequate antimicrobial concentrations. (Red Book 2021)





oral

# pulmonary hydatid cyst

مریم رستمیان  $^1$  ©  $^0$ 

استادیار بیماریهای عفونی کودکان دانشگاه علوم پزشکی سمنان ۱

**Abstract**: pulmonary hydatid cyst Dr.maryam rostamyan Cystic echinococcosis is a zoonotic parasitic disease caused by the larval stages of the cestode Echinococcus granulosus.the hydatid cysts tend to form in the liver (50-70%)or lung (20-30% of patient )but may be found in any organ of the body including brain,heart,and bones(



# A case of interrupted aortic arch associated with MoyaMoya syndrome

Alireza Dehestani <sup>1</sup> © P

Abstract: An eight-year-old girl was referred to our hospital with a history of epigastric abdominal pain in the past 2 years with elevated transaminase and gamma-glutamyl transferase in favor of choledochal cyst. MRCP confirmed the type III choledochal cyst. She also had a history of pallor attacks and headache diagnosed as focal seizure that was treated by carbamazepine. Taking blood pressure in the pediatric cardiology clinic showed systolic blood pressure of 200mmHg in the right arm. On physical examination pulse of the lower extremities was weak and had a systolic pressure of 90mmHg with a 110mmHg pressure gradient between the right upper and lower extremities. Echocardiography showed interrupted aortic arch type A and prominent left ventricular hypertrophy. Brain MRI was done that showed cystic changes and malacia due to the previous infarct in the LMCA territory and multiple collaterals in the basal ganglia and recommenced MRA for assessing Moya Moya syndrome and vascular abnormalities. MRA and CT angiography confirmed the diagnosis of interrupted aortic arch type A and isolated left subclavian artery in addition to the hypoplastic distal vertebral artery. Genetic study of familial Mediterranean fever (FMF) was also positive for heterozygote E148Q. She was scheduled for cardiac surgery with insertion of a tube graft between ascending and descending aorta and reimplantation of the left subclavian artery. Moyamoya disease is defined by progressive stenosis of arteries of Willis circles resulting in collateral formation. It presents by ischemic or hemorrhagic stroke, seizure and ischemic attack. Isolated form with neurologic manifestation is the so-called Moyamoya disease but Momamoya syndrome is a vasculopathy secondary to other acquired or congenital diseases including autoimmune diseases such as Takayasu arteritis, systemic lupus erythematosus (SLE), antiphospholipid antibody, Behcet disease, hemolytic uremic syndrome, trisomy 21, atherosclerosis and vascular interruption. After surgery, she had episodes of hypotension and loss of vision that were treated by titration of vasoactive agents such as norepinephrine. Further assessment revealed the involvement of other arterial territories such as iliac arteries that raised suspicion for inflammatory vasculitis such as Takayasu arteritis. She was treated with corticosteroids and DMARDS such as Cellcept.





#### Cardiology

oral

# A Case of Myocarditis Presenting as Myocardial Infarction with Nonobstructive Coronary Arteries (MINOCA) Syndrome

Reza Shabanian <sup>1</sup> © P

Abstract: Myocardial infarction with nonobstructive coronary arteries (MINOCA) is characterized by clinical evidence of myocardial infarction with normal or near normal coronary arteries on angiography. Several pathophysiologic mechanisms have been postulated as the etiology of MINOCA. Here we report an 8-year-old girl with clinical evidence of myocardial infarction. She was admitted to the emergency department with symptoms of palpitation, exertional dyspnea, shortness of breath, nausea and abdominal pain. She had fever and malaise for the past five days. One month before admission, she and her family had positive PCR tests for COVID-19. Physical examination revealed tachycardia with S3 gallop and holosystolic murmur of III/VI at the apex. The blood pressure was 90/50mmHg. Except for mild hepatomegaly, the other systemic examination was normal. Laboratory exam showed a rising level of troponin I, positive IgG antibody for anticardiolipin and borderline FANA test (1/80 IF). Ventricular tachycardia and accelerated idioventricular rhythm were seen in electrocardiographic monitoring. Coronary arteries were patent without any evidence of stenosis or interruption in CT angiography. Myocarditis was diagnosed as the underlying cause of MINOCA. Following IVIG and steroid therapy, normal sinus rhythm was returned and she experienced notable improvement in her cardiac function at follow-up echocardiograms.





## Acute myocarditis in children

دکتر پروین اکبری اسبق $^{1}$  ©  $^{0}$ 

استاد فوق تخصص قلب كودكان ١

Abstract: Background: Myocarditis is inflammation of the myocardium with necrosis of cardiac myocytes. Myocarditis may be caused by many disorders (eg. infection, cardiotoxins, drugs, and systemic disorders such as sarcoidosis) but is often idiopathic. Viral infections are the most common causes of acute myocarditis. Affected children often have a prodrome of fever, malaise, and myalgia. Clinical manifestations of acute myocarditis in children can be nonspecific. Some children may present with easy fatigability, poor appetite, vomiting, abdominal pain, exercise intolerance, respiratory distress/tachypnea, dyspnea at rest, orthopnea, chronic cough with wheezing, chest pain, unexplained tachycardia, hypotension, syncope, and hepatomegaly. Supraventricular arrhythmias, ventricular arrhythmias, and heart block may be present. A subset of patients have fulminant myocarditis and present with cardiovascular collapse, which may progress to severe cardiogenic shock, and even death. A high index of suspicion is crucial to its diagnosis and timely management. Diagnosis is based on symptoms and clinical findings of abnormal electrocardiography (ECG), cardiac biomarkers, and cardiac imaging in the absence of cardiovascular risk factors. Endomyocardial biopsy confirms clinical diagnosis of myocarditis. Treatment depends on the cause, but general measures include drugs to treat heart failure and arrhythmias and rarely surgery (eg, intra-aortic balloon pump, left ventricular assist device, transplantation). Immunosuppression is of use in certain types of myocarditis (eg, hypersensitivity myocarditis, giant cell myocarditis, myocarditis caused by sarcoidosis). Keywords: acute myocarditis, etiology, clinical manifestation, diagnostic tests.





# Cardiac murmur detected at new born examination; we should be cautious



استادیار دانشگاه علوم پزشکی تهران ۱

Abstract: Cardiac murmur detected at new born examination; we should be cautious Toktam Sheykhian: Assistant Professor of pediatric cardiology, Imam Khomeini hospital complex, University of Medical sciences: Tehran. Tehran Iran. E-mail address: toktamsheykhian@gmail.com. Coronary artery fistula (CAF) is abnormal communication between a coronary artery and a cardiac chamber or a great vessel, which are the most common congenital malformation of the coronary arteries. CAF accounting for 0.3% of patients with congenital heart disease. Small fistulas usually do not cause any hemodynamic compromise. However, the larger fistulae can cause coronary artery steal phenomenon, which leads to ischemia of the segment of the myocardium perfused by the coronary artery. Case presentation: A two days old male neonate was referred to our clinic for consultation because of heart murmur. The neonate was term and born by normal delivery from healthy mother. Birth weight and Apgar score were normal. The neonate was asymptomatic. In physical exam (PE), systolic murmur II/VI was heard at LSB. Other PE were normal. ECG and CXR were normal. Echocardiography revealed dilation of coronary arteries (RCA and LAD). Abnormal flow was seen on the end part of ventricular septum and fistula was visible from distal of RCA to right ventricle (RV). After one-month coronary CT angiography confirmed prominent RCA and LAD. The RCA has a fistula connection to the inferior RV apex after receiving branch from distal of LAD. The patient was followed up closely by clinical and echocardiographic evaluation. Conclusion: heart murmurs in the first days of birth can be different from the other periods of life and should not be easily considered as innocent murmur. TTE is a useful method that should be considered in the investigation and follow up of pediatric coronary artery fistula.





## Coarectation misdiagnosis and the importance of positioning of the in infant and childrens in diagnosis



**Abstract**: Coarectation is one of the common cardiovascular diseases of children, and delay in its diagnosis can cause severe and sometimes irrevercible damage of body organs such as kidney, brain, heart, etc. Echocardiogeraphy is the choice of the diagnosis of coarectation and the Suprasternal view is the view of choice. Neck position is very important for 2D and doppler assessment of coarectation diagnosis. High-quality two-dimensional echocardiographic images of CoA can be readily obtained in infants and young children. Localized stenosis just beyond the origin of the left subclavian artery may be visualized in suprasternal views . A "posterior shelf" of fibrocystic tissue protruding from the posterior aspect of the aorta and oriented toward the ductus arteriosus may be seen. Other findings, including isthmus and transverse arch hypoplasia, poststenotic dilation, and delayed systolic peaking with diastolic runoff in the descending aorta pulse-wave Doppler waveforms, are all consistent with significant CoA. Color Doppler interrogation may localize the site of obstruction when twodimensional imaging is difficult or inconclusive. Continuous-wave Doppler interrogation from the suprasternal window will detect a high-flow velocity across the CoA and may aid in determining the hemodynamic severity of obstruction, with a pattern of diastolic runoff. This may be composed of two superimposed signals, representing a low-velocity flow proximal to the CoA, and a higher-velocity flow across the CoA itself.





### Cardiology

oral

### **Convulsive Syncope**

## Reza Shabanian <sup>1</sup> ©, Minoo Dadkhah <sup>2</sup> ®

**Abstract**: Convulsive syncope is characterized by abnormal movements after fainting that is due to the decreased blood flow to the brain. Convulsive episodes may be the clinical presentation of either the epilepsy or the cardiovascular disorders. Here we report a 5-year-old boy who suffered from recurrent episodes of convulsive syncope, thought to be epileptic in origin, however refractory to different antiepileptic drugs. The electrocardiographic monitoring revealed polymorphic ventricular tachycardia after noxious stimulation and epinephrine injection. Genetic study confirmed the mutation of Calsequestrin 2 in favor of the diagnosis catecholaminergic polymorphic ventricular tachycardia. Hence, the differential diagnosis of epilepsy and cardiovascular convulsive syncope can be quite misleading and clinical features may not always be reliable.





### Cardiology

oral

### **Pulmonary hypertension**

## Mohamad-Taghi Majnoon MD <sup>1</sup> © P

### Abstract:

- Pulmonary hypertension (PH) is an important cause of morbidity and
- mortality in pediatric patients
- Pulmonary hypertension has been classically defined as a mean
- pulmonary artery pressure (PAP) greater than 25 mm Hg at cardiac
- catheterization in a resting state. Recently at the 6th World Symposium
- of Pulmonary Hypertension (WSPH), PH was redefined as a mean PAP
- greater than 20 mm Hg due to recognition that normal mean PAP in
- adults is 14.0+/–3.3 and that some patients with "borderline" PH (mean
- PAP 20 to 25 mm Hg)
- PAH describes a subpopulation of patients with PH characterized hemodynamically by the presence of precapillary PH including an end-expiratory pulmonary artery wedge pressure (PAWP) ≤ 15 mm Hg and a pulmonary vascular resistance (PVR) ≥3 Wood units
- The causes of PH have been grouped into five categories;
- each category shares similar features, such as treatment and pathobiology, but there is overlap between groups.





### Pericardial effusion in pediatrics with covid 19 infection



Abstract: There are severeal forms of cardiac involvements in pediatrics who affected with covid 19 include: myocarditis ,endocarditis ,pericarditis ,conduction system disorders , coronary artey ectasia and aneurysm , thromboembolic events and ... Pericarditis is the inflammation of pericardial layer of the heart and is a common type of cardiac involvements in COVID-19 pandemia. Acute pericarditis may present with precordial or substernal chest pain. The pain is described as squeezing, sharp or dull, and characteristically is worse in the supine position. The patient will prefer to sit upright leaning forward, and may refuse to lie down to be examined. The pain worsens with inspiration, coughing, and movement. Younger children may present with atypical symptoms. Respiratory distress is uncommon unless tamponade or pulmonary disease is present. Rarely, abdominal pain can result from hepatic distension in patients with quickly accumulating effusions. Fever may be present. We have many pediatric patients with a specific type of pericardial effusion that include asymmetric and partially localized pericardial effusion that is mostly pre RV .Becasue of its localized and non common type of pericardial effusion it may be missed in rootin echocardiogeraphy .







### The Comparison Between Intravenous Acetaminophen Versus Oral Ibuprofen in Preterm Newborns With Patent Ductus Arteriosus: A Clinical Trial

Behzad Mohammadpour Ahranjani <sup>1</sup> © P, Hosein Dalili <sup>1</sup>, Zeinab Harif Nashtifani <sup>1</sup>, Mamak Shariat <sup>1</sup>, Mohammadrafie Khorgami <sup>1</sup>

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Abstract: - Oral ibuprofen has been known as a conventional treatment for closing patent ductus arteriosus (PDA) in preterm newborns. Since the use of it might lead to various side effects, other treatments needed to be evaluated. Therefore in a prospective study, we compared the efficacy and safety of intravenous acetaminophen versus oral ibuprofen for the closure of PDA. In this study which was done prospectively and under control, 50 preterm neonates with gestational ages and weights less than 37 weeks old and 2500 grams, respectively, who had PDA, large enough hemodynamically, were included in the study. The patients were divided into two groups: A (intravenous acetaminophen) & B (oral ibuprofen). The two groups were given at most two 3-day courses of the medication (the second course if necessary) and evaluated at the end of each course by echocardiography so as to determine the response to the treatment at each step. The rate of ductal closure, the need for additional treatment, side effects, complications and the newborn's clinical status were recorded. The rate of ductal closure in the both groups after one course of treatment was similar and showed no meaningful significance statistically (P=0.306). But that of the side effects was much higher in group B with a P=0.021. Intravenous Acetaminophen is not only as efficacious as oral Ibuprofen for the treatment of PDA in preterm infants, but also is less likely to lead to side effects and complications

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### pulmonology

oral

### Croup

### Seyed Hossein Mirlohi <sup>1</sup> © P

**Abstract**: Croup (laryngotracheitis) is a respiratory illness characterized by inspiratory stridor, barking cough, and hoarseness. It typically occurs in young children (typically between ages six months to three years) and is chiefly caused by parainfluenza virus. Croup is rarely caused by bacterial infection with the exception of Mycoplasma pneumoniae, which can cause a mild croup-like illness CLINICAL PRESENTATION Symptoms usually begin with nasal discharge, congestion, and coryza and progress over 12 to 48 hours to include fever, hoarseness, barking cough, and stridor. Biphasic stridor at rest is a sign of significant upper airway obstruction Croup is usually a self-limited illness, and the cough typically resolves within three days. Other symptoms may persist for seven days Radiographic evaluation of the chest and/or upper trachea is not indicated. Outpatient treatment — For children with mild croup who are seen in the outpatient setting, we suggest a single dose of oral dexamethasone (0.15 to 0.6 mg/kg, maximum dose 16 mg) or oral prednisolone (1 mg/kg) MODERATE TO SEVERE CROUP:— We recommend glucocorticoid therapy for all children with moderate to severe croup. Dexamethasone (0.6 mg/kg, maximum of 16 mg) is generally the preferred glucocorticoid in this setting. Dexamethasone should be administered by the least invasive route possible. Nebulized epinephrine – We recommend nebulized epinephrine in all patients with moderate to severe croup. After three to four hours of observation, children who remain comfortable may be discharged home if they meet the following criteria: •No stridor at rest • Normal pulse oximetry • Good air exchange • Normal color • Normal level of consciousness • Demonstrated ability to tolerate fluids by mouth • Caregivers understand the indications for return to care and would be able to return if necessary





### Evaluation of child with hypoxemia

فاطمه طریقت منفرد  $^1$  فاطمه طریقت منفرد

فوق تخصص ریه کودکان ۱

**Abstract**: Evaluation of child with hypoxemia Signs and symptoms: Patients may be lethargic, irritable, anxious, or unable to concentrate. Children with respiratory distress commonly sit up and lean forward to improve leverage for the accessory muscles and to allow for easy diaphragmatic movement. Children with epiglottitis sit upright with their neck extended and head forward while drooling and breathing through their mouth. The respiratory rate and quality can provide diagnostic information, as exemplified by the following: Bradypnea Tachypnea The patient should also be evaluated for the following: Stridor Wheezing Crackles Decreased breath sounds Paradoxical movement of the chest wall Accessory muscle use and nasal flaring Cardiovascular signs: Tachycardia and hypertension Gallop Bradycardia Diagnosis: ABG CBC Electrolyte abnormalities Alveolar-arterial oxygen difference PaO2/ fractional concentration of inspired oxygen (FiO2): Indicates gas exchange Oxygen index Dead-space volume to tidal gas volume (VD/VT) Intrapulmonary shunt fraction (Qs/Qt) Imaging studies: Common radiographic findings associated with respiratory failure include the following: Focal or diffuse pulmonary disease (eg. pneumonia, acute respiratory distress syndrome [ARDS]) Bilateral hyperinflation (eg, asthma) Asymmetrical lung expansion suggesting a bronchial obstruction Pleural effusion Cardiomegaly Bronchoalveolar lavage and lung biopsy Management: For partial upper-airway obstruction (eg, from anesthesia or acute tonsillitis), place a nasopharyngeal airway to provide a passageway for air. An oropharyngeal airway can be used temporarily in the unconscious patient. For extrathoracic airway obstruction: Inspired humidity Heliox Racemic epinephrine Systemic corticosteroids Nebulized hypertonic (3%) saline Lung and respiratory pump support: Oxygen therapy: Supplemental oxygen is the initial treatment for hypoxemia Humidified high-flow nasal cannula therapy (HHFNC) Continuous positive airway pressure (CPAP) Noninvasive positivepressure ventilation (NPPV) Conventional mechanical ventilation Inverse ratio ventilation Airway pressure release ventilation (APRV) High-frequency oscillatory ventilation (HFOV) Prone positioning Inhaled nitric oxide (NO) Extracorporeal life support (ECLS)





### Management of Foreign body aspiration in children

Alireza Eshghi <sup>1</sup> © P

**Abstract**: Foreign body aspiration is one of the cause of mortality and sudden death in children.It is common at 6 months to three years old and is common in boys than girls. The most common material for aspiration are seeds. before 3 years old both main bronchi are affected equally. after this age trap in right side is more common. The most common history in FBA is choking that is a sudden cough that was not present previously. If history of choking was present the patient needs to bronchoscopy and normal imaging and examination dose not change the management. So history of choking is equall to do bronchoscopy. There is a different between FBA & FBI. In FBI we may have drooling & nausia and vomiting In history. Coins are uncommon for FBA but if a coin was aspirated in trachea it became vertically in anterior posterior CXR because of muscular wall in posterior of trachea. Bronchoscopy can be do either flexible or rigid and both of them use for FBA management. In flexible bronchoscopy we use basket and in rigid bronchoscopy optical or simple instrument is used. After bronchoscopy CXR was done for R/O pneumothorax.





### Parapneumonic effusion and Empyema

### Masoumeh Gasempour Alamdari <sup>1</sup> © P

Abstract: Parapneumonic effusion (PPE), including simple and complicated effusion and empyema, is the most common complication of pneumonia in children. Parapneumonic effusions are pleural fluid collections developing secondary to an adjacent bacterial infiltrate. parapneumonic effusions are exudates (as opposed to transudates) if at least one of the following criteria is fulfilled: (1) pleural fluid lactate dehydrogenase (LDH)/plasma LDH>0.6 of pleural >200 IU/l, (2) pleural fluid protein/plasma protein >0.5, (3) with a glucose levels lower than 60mg/dl. The development of PE is a continuum from clear fluid with low numbers ofwhite blood cells to overt pus, this continuum is subdivided into three stages:1. "Exudative stage" 2. "Fibrinopurulent stage" 3. "Organizational stage". In clinical practice, PE staging is not straightforward. Pleural fluid analysis has long been used to classify pleural effusions(EMPYEMA) based on the following characteristics: high numbers polymorphonuclear leukocytes, LDH> 1,000 IU/l, glucose lower than 30 and PH <7.2. Most clinical studies on PPE are reference hospital-based, and there is currently no consensus on the most appropriate treatment. Small PPE (<10 mm thick) can usually be managed conservatively. Several clinical trials have been carried out to verify the most appropriate method to drain complicated effusion and empyema (CE/E). Conservative treatment of CE/E is based primarily on antibiotics, restricting chest tube pleural drainage (CTPD) or video-assisted thoracoscopy to the most severe or treatment-resistant cases. Chest tube placement with pleural fibrinolytic medication administration has been established as an effective first-line treatment option in pediatric pneumonia with complicated pleural effusion. Indications for chest tube placement include both complicated parapneumonic effusions and empyema. Reported fibrinolytic dosing has been empirical, weight-based or stratified based on pleural US complexity. While planned frequency of reported dosing has been once, twice or three times daily, data on actual pleural doses administered per day during fibrinolytic therapy are lacking in the published literature.





### Pulmonary hydatid cyst in children: A case report

لادن افشار خاص  $^{1}$   $^{\odot}$   $^{\odot}$  نسرین حسینی نژاد  $^{1}$ 

Abstract: Nasrin Hoseiny Nejad. Ladan Afshar Khas. Aliasghar children Hospital. IUMS Pulmonary hydatid cyst in children: A case report We report an eight -year-old girl with chronic cough and respiratory distress. She was suffering of progressive and productive cough for more than one year. She was diagnosed as asthma, partially controlled by asthma medications. She was underweight, and tachypnea, retraction and coarse crackles, wheezing with silence in some parts of chest were revealed. There was no other positive point in physical examination. In chest radiography, there were multiple round mass like lesions (about seven round masses with 4 centimeters of diameter) in different parts of lung surrounded by normal lung parenchyma. There was a haziness (consolidation) in left lower lobe with pleural effusion. In chest CT Scan, lesions were revealed cystic. Liver and other organs were intact. In laboratory exam, Anti-Echinococcus IgG was detected. Albendazole was prescribed and after one-week, excisional surgery was done, which was repeated three times on different parts of lung. Patient's signs and symptoms were gradually declined. After 6 months of treatment by Albendazole, chest CT scan was found normal. In the next visit of follow up, 2 new cysts of 3 and 4 centimeters in diameter were revealed in Left lower lobe and right upper lobe. Albendazole was prescribed for more 6 months. The next imaging showed that cysts were about 2 centimeters of diameter. After a one year of treatment with Albendazole, no more cyst was found and treatment was stopped. During all these times, evaluation of other organs showed no pathology. Conclusion: Pulmonary involvement by ruptured hydatid cysts is difficult to control and recurrent. Therefore, prevention is very important, especially in endemic areas. Furthermore, early diagnosis, before cysts rupture has a significant impact on prognosis. Key Words: Hydatid cyst, Pulmonary, pediatric, Albendazole





### pulmonology

oral

### Spirometry: test and interpretation

### Seyed Javad Sayedi <sup>1</sup> © P

**Abstract**: Spirometry is a physiological test that measures the maximal volume of air that an individual can inspire and expire with maximal effort. The primary signal measured in spirometry is either volume or flow as a function of time. The most relevant measurements are the FVC, which is the volume delivered during an expiration made as forcefully and completely as possible starting from full inspiration, and the FEV1, which is the expiratory volume in the first second of an FVC maneuver. Spirometry enables measuring the effect of a disease on lung function, assessing airway responsiveness, monitoring disease course or the result of therapeutic interventions, assessing preoperative risk, and determining a prognosis for many pulmonary conditions. It is a valuable tool that provides important information to clinicians which is used together with other physical findings, symptoms, and history to reach a diagnosis. Clinically useful spirograms must be acceptable (meet the criteria that comprises a good quality maneuver) and repeatable (the two highest FEV1, FVC and VC from three acceptable maneuvers are in close agreement). A normal spirometry test is characterized by FVC, FEV1, and FEV1/FVC all within the normal range. The flow-volume loop in normal patients will be tall and wide. An obstructive pattern is characterized by FEV1/FVC < lower limit of normal (LLN). Obstructive spirometry pattern is usually, accompanied by FEV1





# Effect of Paracetamol versus Ibuprofen in Adenotonsillectomy

بهروز امیرزرگر  $^1$ , فاطمه میراشرفی  $^{0}$   $^{0}$  سارا رهاوی عز آبادی  $^{1}$ 

Abstract: The present study aimed to compare the effects of paracetamol and ibuprofen on pain, bleeding, nausea, and vomiting following adenotonsillectomy in children. Materials and Methods: This was a prospective, double-blinded, randomized clinical trial. Block randomization was used to assign 50 patients to two groups of paracetamol and ibuprofen. In the paracetamol group, subjects received 15 mg/kg oral paracetamol 30 minutes before the induction of anesthesia, followed by the same dosage every 6 hours postoperatively. Meanwhile, the ibuprofen-treated group took 10 mg/kg oral ibuprofen 30 minutes before and every 6 hours after the operation. The subjects in both groups received the medication for three postoperative days. The postoperative pain score was assessed 6 hours after the surgery and during the second and the third postoperative days. Nausea and vomiting episodes were recorded in the first postoperative day and first postoperative week. Results: Based on the results, intraoperative and postoperative bleeding in both groups was not significantly different. The mean score of pain in the first postoperative day (6 hours after the surgery) and the second and the third postoperative days did not show any statistical difference. The ibuprofen group experienced fewer vomiting episodes, compared to the paracetamol group during the first postoperative day (P=0.011). Vomiting episodes in the first postoperative week did not illustrate any significant difference. Conclusion: As evidenced by the results of the current study, Ibuprofen had the same effect on the alleviation of postoperative pain, caused fewer vomiting episodes, and did not cause excessive bleeding as an NSAID. Therefore, oral administration of ibuprofen is suggested for pain relief and management of other complications following adenotonsillectomy in children.





### **Inguinal Hernia in Children**

مريم قوامي عادل  $^1$  ©  $^{\mathbb{P}}$ 

دانشگاه علوم پزشکی تهران-گروه جراحی کودکان ۱

Abstract: Inguinal hernia is one of the most common pediatric surgical presentations. The incidence of inguinal hernias is approximately 3% to 5% in term infants and 13% in infants born at less than 33 weeks of gestational age and it is six times more common in boys. Over 99% of inguinal hernias in children are indirect and a direct hernia is extremely rare in Children. Patients usually present once a parent has noticed a lump or swelling in the groin. Hernias can be classified as reducible or irreducible. If the hernia is irreducible, it is important to distinct between strangulated and non-strangulated hernias. The child should be examined with warm hands and in a comfortable environment. Hydrocoeles, undescended testis, chordal cyst, inguinal lymphadenopathy, Idiopathic scrotal edema, abscess in the inguinal region and less commonly varicoceles and testicular tumors are the differential diagnosises. Management-Inguinal hernias in both term and preterm infants are commonly repaired shortly after diagnosis to avoid incarceration of the hernia. Infants aged





## Middle Ear Status in Cleft Lip and Palate Patients: A Five-Year Follow-Up.

Fatemeh Mirashrafi <sup>1</sup> © P, Sara Rahavi-Ezabadi <sup>1</sup>

**Abstract**: Introduction: The best strategy to treat otitis media with effusion in cleft lip/palate patients is still under debate. This research aimed to evaluate the otologic outcomes in children at least five years post-repair. Materials and Methods: A retrospective study was conducted on 40 children who underwent palatoplasty between January 1, 2012, and January 1, 2014, at Children's Medical Center (Tehran, Iran). Patients had intervelar veloplasty under magnification (Sommerlad's Technique). Based on patients' charts, their age, gender, cleft type, date of palatoplasty, as well as the date and the frequency of ventilation tube (VT) insertion, were recorded. Furthermore, otomicroscopy, middle ear status, and tympanometry were assessed five years postoperatively. Results: There was no significant difference in middle ear status between children with complete and incomplete cleft palates. The mean age at the time of study and the mean follow-up duration were significantly higher in the normal middle ear group, compared to the abnormal middle ear group (7.7±1.6 vs. 6.8±0.9, P=0.03 and 6±1.15 vs. 5.42±0.9, P=0.04, respectively). Middle ear status was not significantly different between early or late palatoplasty patients. In addition, the frequency and timing of VT insertion were not significantly different between the two groups. Conclusions: Middle ear status improved as patients grew older; however, the age of palatoplasty and the frequency of VT insertion were not significant prognostic factors in patients who underwent intervelar veloplasty under magnification.





### retinopathy of prematurity

## افسر دستجانی فراهانی $^1$ $^{f C}$

مرکز رتینوپاتی بیمارستان فارابی،مرکز تحقیقات چشم پزشکی ترجمانی ا

**Abstract**: Pathogenesis: Retinopathy of prematurity is a potentially blinding, preventable and treatable disease. It happens in premature neonates. Normal vascularization in nasal side will be complete at 36 PCA and 40 in temporal part. In premature delivery retinal vascularization is incomplete and its severity depends to severity of prematurity. The disease has 2 important phases: First phase happens 30-32 weeks post conception age. Hyperoxia exposure lead to decrease VEGF level and then decreased vessels growth and shedding of previous vessels, concomitantly decrease of IGF1 level lead to decrease vessel growth. These 2 events induced hypoxia. Second phase of RoP starts from 31-44 weeks post conception. In this phase the hypoxic avascular Retina produces massive VEGF and then Neovascularization and progression of abnormal vessel growth happens in this phase. Clinical course: RoP has 5 stages. 1-A demarcation line between vascular and avascular area. 2- A wider and deeper ridge between vascular and avascular area. 3-Neovascularization Tufts 4-partialy detached retina 5totally detached retina How can we prevent blindness and treat RoP? With 4W and 2 H: Who: Iranian national guideline suggests Gestational age less than 34 weeks and or birth weight less than 2001 gram. If we follow the American guideline 8.4% of premature neonates who needs treatment will lost. In cases with 37>GA

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### Algorythms

oral

## Review some mistakes in pediatric imaging

## Seyed Mehdi Alehossein <sup>1</sup> © ®

هیئت علمی دانشگاه علوم پزشکی تهران ۱

**Abstract**: In this lecture I present some cases with routine scenarios of ill children that were referred to radiology department and requested for taking emergent radiography, sonography, CT scan or MRI. There are many pitfalls that clinicians and radiologists should be aware of when and how request these studies and interpreting the findings. Surely there are some inherent mistakes in taking history, physical exam, imaging technique and finally obtained conclusion. We discuss some sampled real cases for the audience with step by step imaging approach and how to avoid mistakes.

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### Teaching the social sciences in Residency

رضا رضایی طاهری  $^1$   $^{\circ}$ 

, 00000

**Abstract**: The social sciences are used to study human society, social organisations, and other phenomena .the term "socials cience" is used to contrast to the so called natural sciences (such as biology, physics, chemistry, and mathematics and their subfields) and encompasses a vast array of disciplines, including sociology, economics, history, political science, and geography. Proposals to integrate social science content to medical education have a long history. As early as 1936, medical historian Henry Sigerist eloquently called on medical students to aquire some knowledge in the social sciences:"not to make you experts in the subject but you must have a certain knowledge in order to be able to understand the world in which you live." A 2004 report by the Institute of Medical (IOM) highlighted the importance of social and behavioural factors in health, described the history of approaches to incorporating social science content into curricula, and recommended curricula topics and strategies for the incorporation of these topics to enhance the social science education provided during medical school. In England, the Behavioural and Social Sciences Teaching in Medicine network described the evolution of behavioural and social science education in British medical education in 2010 report funded by the Higher Education Academy. A contemporary rationale provided for the incorporation of the behavioural and social sciences teaching in medical education is the need for competency framework to ensure physicans are trained in all skills necessary to improve and maintain patient's health. While the question of whether Competency-Based Medical Education will achieve its aims remain to be answered, the current context of medical education in many countries requires educators and programme directors to work within the CBME paradigm. Accordingly, education in the biomedical sciences, the traditional domain of medical knowledge, may not afford medical learners a foundation to support the full suite of competency expected of physicans





# Investigation of Follow-up Results of Patients with Primary Immunodeficiency Diagnosed in Yazd province during 2008 and 2018

Marzie Vaghefi <sup>1</sup> © P, Nasrin Behniafard <sup>1</sup>, Mohammad Hasan Bemanian <sup>1</sup>

Iran

**Abstract**: Background: Primary immunodeficiency diseases are a group of disorders that occur as a result of primary disturbances of the immune system and make a person susceptible to recurrent infections. No detailed study has been conducted so far in Yazd province on the prevalence, effective factors, and follow-up of patients with immunodeficiency; on the other hand, there have been recent advances in treatment including bone marrow transplantation to help improve the quality of life of these patients. Thus, this study investigated this issue. Materials & Methods: In this historical cohort study, census method was used to select and assess all the patients with primary immunodeficiency diagnosed by a subspecialist of asthma, allergy, and clinical immunology and approved by a specialist on the basis of patient interview, examinations, Para clinical tests, and genetic investigation during 2008-2018. Finally, the gleaned data were analyzed by SPSS21. Results: The results of the present study showed that the most common types of immunodeficiency in the study population were "predominantly antibody deficiencies" (36.1%) and the most common diagnoses were CVID, SCID and CGD with frequencies of 20.6%, 11.3% and 7.2%, respectively. Of the patients studied, 62 were boys and 35 were girls. The mean age at diagnosis, age of onset of symptoms, and delay in diagnosis in individuals with immunodeficiency were 8.14, 3.88 and 4.26 years, respectively. The total mean survival of patients with primary immunodeficiency was 14.95 years, their 5-year survival was 82%, and 10-year survival was 75%. Conclusion: it is suggested that family screening be performed as far as possible on high-risk children with recurrent infections, consanguineous marriages, and family history of immunodeficiency to take effective steps for prompt diagnosis and control of the disease. Keywords: Primary immunodeficiency, survival, Yazd province, historical cohort.





# Evaluation of the relationship between neonatal convulsive seizure semiologies and underlying etiologies in neonates hospitalized in childrens' medical center between years 1394-1399

# Sevil Abdolmohamadian <sup>1</sup> © P, Reza Shervin Badv <sup>2</sup>, Ziba Mosayebi <sup>3</sup>, Mahmoud Mohammadi <sup>4</sup>, Mahmoudreza Ashrafi <sup>4</sup>, Kayvan Mirnia <sup>5</sup>

### Sciences

**Abstract**: Seizures are the most common neurological emergency in the neonatal period. The aim of our study was to evaluate the relationship between seizure semiology and etiological factors in our population of neonates being hospitalized in children's medical center NICU between years 1394-99, trying to find out whether specific kinds of clinical presentations are related to specific etiologies. We selected all the neonates(age 1–28days for term newborns, and up to 44weeks of PMA for preterm neonates) presenting convulsive seizures. Seizures were divided into: Automatisms, Clonic, Epileptic spasms, Myoclonic, Tonic, Autonomic and Sequential Seizure Type. In order to classify seizure semiology, we evaluated the first clinical sign and not the predominant feature, according to the new approach to seizure classification proposed by ILAE in 2017 we divided etiologies into 7 major categories: Vascular, infections, HIE, genetic, metabolic, structural, unknown. This study included 131 patients [66 males, 65 females], with mean gestational age of 37.7±2 weeks (ranging between 30.0 and 43.0 weeks). Only 17% of patients were born premature. 73% of patients were born by C/S. Seizure onset ranged between 1 and 70 days (mean 6 days). 83.2% of patients had birth weight of 2500-4000 gr, 5.3% were born heavier than 4000 gr, 10.7% had LBWs, only one patients had ELBW. Mean birth weight was 3100±599 gr. Most common seizure semiology was Myoclonic(43.5%) and least common semiologies were Autonomic & Sequential(both 1.5%). Most common seizure etiology was Unknown Etiologies(64.1%) and least common etiology was Vascular Etiologies (3.1%). The current reality in many regions of the world is that access to even the most basic EEG studies is not possible. This can be used to lobby for better facilities even if the process is challenging and takes many years to achieve. This study did not find any relationship between seizure semiology and parameters like age, gender, gestational age and delivery route.



### gene therapy in duchenne muscular dystrophy

# Seyyedeh Azade Hoseini Nouri <sup>1</sup> © ®, Mohamadmehdi Karambin <sup>1</sup>, Yasaman Ashjari <sup>1</sup>

Abstract: Gene therapy in duchenne muscular dystrophy Seyyedeh Azade Hoseini Nouri1, Mohammad Mehdi Karambin1, yasaman ashjari1 1. Pediatric Disease Research Center, Guilan University of Medical Sciences, Rasht, Iran. Abstract: Introduction: Duchenne muscular dystrophy (DMD) is a fatal X linked genetic disease affecting male children and is caused by a mutation in the gene encoding for dystrophin. Dystrophin is a structural protein that maintains the integrity of muscle fibres and protects them from contraction-induced damage. This mutation leads to progressive muscle deterioration and causes cardiac and respiratory difficulties. Patient will be wheelchair bound until 12 year old and few patients surviving beyond their third decade of life. There is no definit cure for this disease. The mutations responsible for Duchenne muscular dystrophy mainly include exon deletions (70% of patients) and point mutations (about 30% of patients) of XP21. The application of gene therapy for the correction of different mutations found in the DMD gene led to the development of several potential therapeutic approaches .A promising approach for treating this lifethreatening disease is gene transfer to restore dystrophin expression using a safe and single infusion, non-pathogenic viral vector called adeno-associated viral (AAV) vector. Gene therapy leads to production of a shortened form of the dystrophin. The CRISPR-Cas 9 (clustered regularly interspaced short palindromic repeat, CRISPR-associated) technology is becoming increasingly precise and is acquiring diverse functions through novel innovations such as base editing and prime editing in animal models. Few of these therapies focus on treating mutations arising in the N-terminal encoding region of the dystrophin gene. Three clinical trials are currently ongoing in the United States by Sarepta Therapeutics, Pfizer and Solid Biosciences, each with a slightly different mini-micro-dystrophin constructs delivered using AAVs of differing serotype. It is worth noting that FDA has imposed a clinical hold on Pfizer's minidystrophin gene therapy candidate PF-06939926 due to the death of a male after





## Intermittent Altered level of Consciousness as a Manifestation of Corona virus infection in Children

## Ladan Afsharkhas <sup>1</sup> ©, Nasrin Hoseiny Nejad <sup>2</sup> ®

Abstract: Intermittent Altered level of Consciousness as a Manifestation of Corona virus Infection in Children Dr ladan Afsharkhas 1, Nasrin Hoseiny Nejad 2 1. Associate Professor of pediatric Neurology, IUMS, Tehran, Iran 2. Associate **Professor** of pediatric Pulmonology, IUMS, Tehran, Iran Back ground: It has known that some children with COVID-19 infection may experience neurologic complications. The aim of this study was to present a child with corona virus infection and central nervous system manifestation. Case:Patient was a 4.5 year old male who was admitted with abnormal gait and intermittent loss of consciousness in our centre. His parents were consanguineous and he was born through an uneventful normal vaginal delivery. He had already normal developmental status. Vaccination was done as routine. He had gastroenteritis one month before hospitalization. He gradually became irritable and aggressive. In neurologic examination, he did not have eye fixation and following although eye examination was in normal limits. He lost swallowing, sitting and walking abilities. Deep tendon reflexes was normal and babinski sing was not detected bilateraly. Signs and symptoms had wax and wane. Brain magnetic resonance imaging (MRI) was performed. Lumbar puncture was down and specimens for evaluation of biochemistry, culture, herpes virus, Epstein bare virus, Cyto-megalovirus and autoimmune encephalitis was sent to a central diagnostic laboratory. Because of pandemic, polymerase chain reaction test for corona virus was checked in nasopharynx specimen. Brain MRI showed some abnormal signals in bilateral thalamus and brain stem in T2 images. Intravenous methylprednisolone pulse therapy was started with probable diagnosis of autoimmune encephalitis. Covid-19 test was positive. Autoimmune panel and virology evaluation in CSF was negative. All manifestations were subsided a few days after treatment. Conclusion: During covid-19 pandemic, some non specific neurologic presentations may occur. In our case, the patient had a dramatically response to corticosteroid. It is recommended to try immune therapy in such patients. Key words: CNS, COVID -19, Immune Therapy





## Sleep habits and related factors in exceptional primary school students

### Saideh Sadat Mortazavi <sup>1</sup> © P, Zahra Mortazavi <sup>2</sup>

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**Abstract**: Background: Sleep is one of the areas of occupation humans and is an effective item in a child's growth and development. Sleep habits of a person form in childhood. The aim of this study is to determine the sleep habits of special students in primary school, Hamadan, Iran. Methods: In this correlational descriptive study, 102 students aged 7-10 years, were selected using the available sampling method in Hamadan city in 2021. Data were collected through a demographic information questionnaire and the Children's Sleep Habits Questionnaire (CSHQ). The questionnaires were completed as a self-report by the parents (mother or father). Data were analyzed using descriptive statistics methods and an independent t-test. Findings: The mean score of the students' sleep habits was in girls  $76.87 \pm 17.24$  and in boys  $73.99 \pm$ 17.99. Sleep duration in girls is  $9.42~9\pm1.40$  hours and sleep duration in boys was  $9.53\pm1.22$ hours, the highest mean score of sleep habits was related to cerebral palsy students. Discussion& Conclusion: The findings of this study showed that sleep habits disorders in special students are prevalent. Keywords: Sleep; Students with special needs; habit sleep References: 1. Choi E, Jung E, Van Riper M, Lee Y. Sleep problems in Korean children with Down syndrome and parental quality of life. Journal of Intellectual Disability Research. 2019;63(11):1346-58. 2. 2. Shamsaei F, Ahmadinia H, Seif M, Khalili A. Sleep Habits of Primary School Students of Nahavand City From the Point of View of Parents. Oom University of Medical Sciences Journal. 2018;12(8):78-85. 3. Agathão BT, Lopes CS, Cunha DB, Sichieri R. Gender differences in the impact of sleep duration on common mental disorders in school students. BMC Public Health. 2020;20(1):148. 4. Hershner S. Sleep and academic performance: measuring the impact of sleep. Current Opinion in Behavioral Sciences. 2020;33:51-6.





# Survey of the health literacy level of parents of children with special needs (3-6 years)

# Saideh Sadat Mortazavi <sup>1</sup> ©, Elham Khadem Hamzeie <sup>2</sup> ®, Zahra Mortazavi <sup>3</sup>

Hamadan, Iran

**Abstract**: Background: The health literacy of parents of children with special needs reflects their skills and ability to receive and apply useful knowledge. The purpose of this study was to determine the level of health literacy of parents of children with special needs (3-6 years). methods: In this descriptive-analytical cross-sectional study, 92parents (64 mothers and 28 fathers), who have children referring to occupational therapy and speech therapy centers, were randomly selected. Data gathering was conducted by demographic and parent health literacy questionnaires, and SPSS software version 16 all data was used to analyze data by independent t-test, ANOVA, Pearson correlation coefficient at a significance level of 0.05. Results: The mean of father and mother's health literacy were  $55.18\pm7.59$  and  $61.72\pm13.56$ , respectively. A significant difference was observed in parents' health literacy, parent's gender (p = 0.019), parent's education level (p = 0.05), and father's occupation. Pearson's correlation coefficient analyses showed a significant negative correlation between parental health literacy and father age (r = -.345, p < .01), maternal age (r = -.418, p < .01), and parental health literacy positively correlated with the number of OT (r = .238, p < .05) and ST Sessions (r = .468, p < .01). Conclusion: parent's health literacy was inadequate, and level of health literacy was higher in younger and more educated mothers and significantly associated with further follow-up of rehabilitation (occupational therapy and speech therapy). It is necessary to plan educational strategies to increase the level of health literacy in parents who have children with special needs, because of these children are vulnerable population. Keywords: Parents Health Literacy, rehabilitation, children with special needs.





### Neonatology

poster

# Evaluation of hyperbilirubinemia and etiology in neonates in Children's Medical Center and Ziaeian hospital from May 2020 to September 2022

Golnaz Ghazizadeh Esslami <sup>1</sup> © P, Mohammad Ali Vazir <sup>r</sup>, DDDDDD DDDDD <sup>r</sup>

**Abstract**: Background: Detection of etiologies and related factors for the development of icterus is crucial to improve the prognosis and decrease the complications in neonates by using early interventions like phototherapy. Due to the lack of sufficient data, the researchers in this cross-sectional study sought to investigate the causes of jaundice among neonates referred to Children's Medical Center and Ziaeian Hospital from May 2020 to September 2022. Methods: 219 neonates entered the study. Variables included gender, age, labor type, maternal diabetes, ABO or RH set up, G6PD, RBC morphology, etc. Statistical analysis was performed using the statistical package for SPSS software version 22. P value





# The association between vitamin D levels and necrotizing enterocolitis in preterm neonates

Kamyar Kamrani <sup>1</sup> © P, Mohammad Reza Zarkesh <sup>2</sup>

عضو هیئت علمی دانشگاه علوم پزشکی تهران – بیمارستان کودکان بهرامی <sup>۱</sup> عضو هیئت علمی دانشگاه علوم پزشکی تهران – بیمارستان محب یاس <sup>۲</sup>

Abstract: Backgroung & purpose: Necrotizing enterocolitis (NEC) with high morbidity and mortality rates is a frequent gastrointestinal disease among preterm infants. This study was conducted to evaluate any relationships between maternal/ neonatal serum vitamin D concentrations and the incidence of necrotizing enterocolitis in preterm newborns, Methods: A prospective case-control study was carried out in an Iranian hospital in 2018. Patients and methods: Thirty-two NICU hospitalized neonates due to NEC and 32 hospitalized neonates due to prematurity with their mothers were considered as the case and control groups. Immediately after delivery, 5 ml of the mother's blood was collected and sent to the laboratory. Two ml of neonate's blood was also collected in the time of admission and sent to the laboratory. Our primary objective was to assess the association between maternal/neonatal vitamin D serum concentrations and the risk of NEC. Results: The means of maternal and neonatal serum vitamin D were  $35.00 \pm 15.94$  and  $33.29 \pm 14.96$ . There was a significant positive correlation between maternal and neonatal vitamin D status (p=0.0001). There were significant associations between NEC and some neonatal factors including neonate's low birth weight (p=0.01), head circumference (p=0.02), and height (p=0.03), as well as low Apgar score at first minute (p=0.04). No significant associations were observed between NEC with maternal and neonatal levels of vitamin D status. Conclusion: Our results showed a significant positive correlation between maternal and neonatal vitamin D status. Although some neonatal characteristics were significantly correlated to NEC, this significant association was not observed with maternal/neonatal levels of vitamin D.





## The parallel paradox of birth and death, a systematic review on perinatal death consequences

## Leila Taheri <sup>1</sup> © P, Zahra Kaffash <sup>2</sup>, Roya Taheri <sup>3</sup>, Zohre Khalajinia <sup>4</sup>

midwifery, Qom University of Medical Sciences. Qom, Iran.

University of Medical Sciences. Department of Midwifery, School of nursing and midwifery, Oom University of Medical Sciences. Qom, Iran.

**Abstract**: Introduction: The death of a baby during pregnancy or immediately after birth is a devastating consequence for families and is still a public health priority in the world. Parents experience psychological, physical, social, and spiritual outcomes. It represents a significant loss experience that can cause acute and prolonged grief reactions. Material and methods: A systematic search was conducted in The PubMed, CINAHL, Scopus and google scholar for eligible publications in accordance with the PRISMA guidelines. The review proposal was registered with the PROSPERO. The quality assessment was done according to JBI Critical appraisal tools. Results: From the identified 1032 potentially relevant studies published from 2012 till 2022, eventually 68 articles met the predetermined inclusion and exclusion criteria. The findings demonstrated that perinatal death is intensely painful and traumatic for many parents and associated with substantial direct and indirect, psychological and social costs to women, families and society. Parents experience pregnancy loss often suffer from feelings of guilt, a sense of failure, depression, social withdrawal, and psychosomatic illness. The results suggest higher levels of stigmatisation are experienced by first-time mothers; who had a mental health diagnosis prior to, or following, their stillbirth; Some research findings reinforce the need for appropriate use of language in health care encounters that acknowledge infant loss and related hopes and dreams and prepares parents for the labor and birth experience. Conclusion: Pregnancy is considered a unique time in life. Although this period is often accompanied by indescribable joy, when women experience complications or miscarriage, stillbirth or infant death, pregnancy can have devastating physical and emotional effects on individuals. Therefore, it is important that medical professionals who care for these families are aware of what they can do to mitigate psychological symptoms among these individuals. Key words: Perinatal Death, Stillbirth, family, Systematic Review







## A case report of hemolytic uremic syndrome in a 13month infant

زینب فدائی  $^2$  و طیبه رضائی  $^2$  و طیبه رضائی  $^3$ 

دانشگاه علوم پزشکی تبریز، بیمارستان آیت ا... حجت کوه کمری مرند، متخصص کودکان و نوزادان <sup>۱</sup> دانشگاه علوم پزشکی تبریز، بیمارستان آیت ا... حجت کوه کمری مرند، سوپروایزر آموزشی <sup>۲</sup> دانشگاه علوم پزشکی تبریز، بیمارستان آیت ا... حجت کوه کمری مرند، دکترای پرستاری <sup>۳</sup>

**Abstract**: Introduction: Hemolytic uremic syndrome is characterized by the triad of hemolytic anemia, thrombocytopenia and acute renal failure. In most parts of the world, this syndrome is mentioned as the most common cause of acute renal failure in children under 4 years of age, which can be seen in two classical and non-classical forms, the classical form of which is followed by diarrhea and the cause of which is verotoxin. Case report: The patient, a 13month-old infant, came to the hospital with symptoms of fever and watery diarrhea, followed by bloody diarrhea, vomiting, and lethargy, and was admitted with the diagnosis of gastroenteritis. Two days after hospitalization, the child has tachycardia, paleness and respiratory distress. A repeat CBC test is requested. The test showed that hemoglobin has dropped from 11 to 7.5 and platelet drop (19000). The Pediatrician immediately sends the patient to an equipped center with a possible diagnosis of HUS. According to the increase of Cr and Urea in the tests, the diagnosis of the patient is confirmed and peritoneal dialysis is started for the child. The child is also injected with Packcell. The infant is discharged after one month of hospitalization and medical procedures in good general condition. Conclusion: Hemolytic uremic syndrome is one of the important diseases of children's medicine, which unfortunately can cause severe and fatal complications. This disease can be misdiagnosed with other diagnoses such as dysentery, parasitic infection, sepsis, and gastroenteritis, and delay in the treatment process. Therefore, in the visit of children, clinical observations and diagnostic tests are very important and require careful examination. Key words: hemolytic uremic syndrome, infant, disease





## Access challenges in children requiring hemodialysis: a review study

فاطمه بهرام نژاد  $\mathbb{C}$  و ناطمه بهرام نژاد  $^2$ 

دانشجوی کارشناسی ارشد پرستاری مراقبت های ویژه دانشکده پرستاری و مامایی علوم پزشکی تهران <sup>۱</sup> تهران، دانشگاه علوم پزشکی تهران استادیار، دانشکده پرستاری و مامایی <sup>۲</sup>

**Abstract**: Introduction: The latest studies of the latest incidence of end-stage renal disease in children (age 0 to 19 years) have grown in two decades. The past approaches for kidney replacement are considered by therapists. Hemodialysis is one of the important topics that, despite its benefits, has many challenges, including challenges in vascular access. This review study was conducted with the aim of reviewing these challenges. Methodology: present review study in 1401 and by searching articles with keywords such as Hemodialysis, children, access to disease, end-stage disease, complications, were performed in the scientific databases of Embis, PubMed, and Scopus. 23 studies of inclusion criteria were included in the present study. Finally, after removing duplicate articles or articles with low quality, 8 studies were included in this study. Results: In children, due to the unique characteristics of this population, such as the usual smaller body and small-diameter blood vessels present distinct challenges for longterm use creates. A central venous catheter is currently the most widely used method of access. A review of several research studies and reviews of fistula or arteriovenous graft as an option it is better to advise. But in children, they become obstructions or fistulas exceptionally small children (weighing less than 10 centimeter) may take. Conclusion: The choice of treatment in children based on the characteristics of the patient, hemodynamic status and personal resources, the duration and possible frequency of hemodialysis, the rate of progression of chronic kidney disease and the patient's health for transfer to peritoneal dialysis can determine the factors that are related to the results. Hemodialysis slows down in pediatric patients and trying to improve them, considering the longer life expectancy, is a significant consideration. This gives high priority to interventions that increase survival and preserve vascular access options for future use. Keywords: hemodialysis, children, vascular





# Gyrate Atrophy presenting in neonatal period: A Case Report.

Shirin Moarefian <sup>1</sup> © P, Ali Rahmanifar <sup>2</sup>, Talieh Zaman <sup>3</sup>

Abstract: Background: Gyrate atrophy is a rare autosomal recessive inherited metabolic disorder affecting visual system caused by mutations in OAT gene resulting in B6 dependent enzyme ornithine amino acid transferase deficiency and hyperornithinemia. The onset is usually in childhood but also can present at birth with hyperammonemia. Here we describe neonatal onset of gyrate atrophy. Case Study: A 7 days female newborn, 2nd child of consanguineous parents whose 1st was deceased neonatal without diagnosis, referred for hypotonia and vomiting. Born by uneventful cesarean section at 36th week with Body weight: 2500 gr and Head circumference: 34 cm. Metabolic work up were normal except hyper ammonemia ,discharged well with sodium benzoate and L-carnitine at 21st days. Normal growth and development but mild myopia at 5 years corrected with glasses. Follow up revealed slightly high ornithine(303±100µmol/L; Normal:20-135) ,so B6 was added to the regimen. Unfortunately at 6 years old parents discontinued treatment and follow up and at 8 years showed night blindness, worsening myopia and gyrate atrophy. So parents again started treatment. Result: She is now 11 years mentally and physically normal improved visually and disappearing eye blindness after low arginine diet ,B6 ,sodium benzoate, L-carnitine and close follow up to normalize ammonia and ornithine. Genetic study showed homozygote mutation in OAT gene: c.290T>C (p.Ile97Thr) with carrier state detection in parents. Conclusion: Gyrate atrophy is a disease of childhood which may present at neonatal period can be prevented if diagnosed genetically at this period of life to differentiate from other urea cycle defects.







### **Endocrinology**

poster

### pharmacotherapy in pediatric obesity

## Mohammad Mehdi Karambin <sup>1</sup> © P, Seyyedeh Azade Hoseini Nouri <sup>1</sup>, Yasaman Ashjari <sup>1</sup>

Abstract: Pharmacotherapy in pediatric obesity Mohammad mehdi Karambin1, Seyyedeh Azade Hoseini Nouri1, yasaman ashjari1 1.Pediatric Disease Research Center, Guilan University of Medical Sciences, Rasht, Iran. Abstract; Introduction; Over the past three decades, the prevalence of overweight and obesity in children and adolescents has increased up to 3 times. It is a multi-systemic medical problem affecting all socioeconomic groups and is associated with an increased risk of other severe comorbidities. Classification of body mass index status in children is based on sex and age-specific CDC or WHO curves. BMI less than %5,%5 -%85, %85 - %95, and >%95 are mentioned as underweight, normal weight, overweight, and obese, respectively. Assessment for obesity risk (dietary habits, eating patterns, daily physical activity, duration and quality of nocturnal sleep) is mandatory during routine well child care. Changes in lifestyle and eating habits of the child are the main principles for gradual weight loss. Changes in lifestyle and eating habits of the child are the main principles for gradual weight loss. Observing proper dietary habits and exercise and nutrition counselling should continue. Initial screen for comorbidities, earlier follow up and Specific behavioral and eating patterns modification for at least 6 month, is recommended if BMI is >85 % or is rising sharply. In adolescents with severe or refractory obesity, pharmacotherapy or surgery is considered after 6 months. Pharmacotherapy, can only be offered for the treatment of obese children after failure of the diet and lifestyle changes, and if the potential benefits outweigh the risks, especially in severe obesity with cardiometabolic, hepatic or respiratory disorders. It is noteworthy that in the case of ignoring behavioral modifications, more significant weight gain may have occurred after discontinuation of pharmacotherapy. Even with drug continuation, weight loss stops around 6 to 9 months after treatment. Losing weight >2kg in the first month, and > 4-5 % baseline weight between





## A Novel Autosomal Recessive FECH Mutation in an Erythropoietic Protoporphyria Patient With Liver Injury: A Case Report with Depiction of Liver Histopathological Features

Fatemeh Elham Mahjoub <sup>1</sup> © P, Zohreh Kavemanesh <sup>2</sup>, Pooria Asili <sup>3</sup>, Maryam Mirahmad <sup>3</sup>, Zahra Hadipour <sup>4</sup>

### Sciences, Tehran, Iran

Abstract: Background: Porphyrias are defects in one of the eight stages of heme synthesis, which are mostly inherited. The accumulation of pathway precursors due to the enzyme deficiency results in different manifestations. Defect in the last enzyme (ferrochelatase) results in Erythropoietic Protoporphyria in which the protoporphyrin-IX is aggregated. The common manifestation is cutaneous photosensitivity; however, liver involvement is the most important one. Case presentation: We report a 2.5-year-old girl who was presented with a history of photosensitivity from early in life. Her laboratory tests and pathological examination revealed cholestatic liver injury with conjugated hyperbilirubinemia in favor of EPP disease. The EPP diagnosis was confirmed by genetic analysis, which demonstrated autosomal recessive ferrochelatase (FECH) gene mutation. Conclusions: The greatest risk for Erythropoietic Protoporphyria patients is liver injury, which must be followed up regularly. The genetic analysis revealed a homozygous likely pathogenic variant (c.743T>C) in the FECH gene with autosomal recessive inheritance, associated with Erythropoietic Protoporphyria. To the best of our knowledge, this variant has never been reported before in the literature.





## Underlying diseases among children hospitalized with COVID-19 in Shahid Sadoughi Hospital, Yazd, Iran

Zahra Nafei <sup>1</sup> ©, Marzie Vaghefi <sup>1</sup> ®, Mehran Karimi <sup>1</sup>

Abstract: Introduction: The 2019 coronavirus disease (COVID-19) outbreak, caused by the novel severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), raised a global health crisis. (1,2) Several studies on this disease have been reported in children, and the differences in infection rates, symptoms, and mortality have been different compared to adults (3,4). The prognosis of COVID-19 is mainly good in pediatric patients without underlying disease but may be worse in cancer patients, especially in people who have recently undergone chemotherapy, radiotherapy, or immunotherapy; it creates a greater risk, which leads to the deterioration of the patient's condition (5) the presence of underlying diseases could increase the mortality rate in children with COVID-19 in the hospital (1) Methods: This retrospective cohort study was conducted among 191 children with COVID-19 hospitalized from March 2020 to July 2021 in Shahid Sadoughi Hospital, Yazd, Iran. Result: In this study, among 191 patients with covid-19, 81 (42.4%) had an underlying disease. The most common underlying diseases were Neurologic (13.1%), Malignancy (7.9%), and Hematologic (4.7%) disease. The mean age of the patients was 7.4 years. Positive PCR tests have been reported in 49 patients (60.5%) (P: 0.014), and 39.5% of patients were admitted to PICU. (P: 0.001) The patients' mean hospitalization duration was five days (1-28 days). There was no significant relationship between the underlying disease with the outcome (p: 0.074) and the severity of COVID-19 (P: 0.99), but in the analysis, a significant relationship was found between the type of underlying disease and the severity of it (P: 0.02), so that patients with heart problems are more likely to have severe and critical conditions. Conclusion: The most common underlying diseases were neurological, malignancy, and hematological. However, patients who had heart disease experienced severe and critical COVID-19. Key words: COVID-19, children, underlying disease





## Iterative Reanalysis of Genome Sequencing: Effective Diagnostic plan for Hypertrophic Cardiomyopathy

Sima Binaafar <sup>1</sup> P, Nejat Mahdieh <sup>2</sup> O

Abstract: Hypertrophic cardiomyopathy (HCM), a dominant disease with delayed penetrance and variable expressivity, affect 1 in 500 individuals worldwide. Since 1989, when MYH7 gene was introduced for the first time, we encounter of rapidly growing reported disease-causing variants. The advent of next-generation sequencing (NGS) has expended robustly the identification of disease-causing genes and variants of HCM. NGS, best-practice technologies, has progressively led to screening of genes associated with HCM mimics. Although genome sequencing is increasingly available in clinical, it is also important to provide a clinically useful reports. In addition to the incidental or secondary findings, the diagnosis of cases with "variant of uncertain significance" (VUS) is a notable challenging status. Clear genotypic information may guide cardiac interventions such as implantable cardioverter—defibrillator placement administration. Fortunately, rapidly growing molecular knowledge represent a profound impact on the assessment of NGS data. This development has associated with databank progress that bring multiple classification changes. Therefore, iterative reanalysis of data of different sequencing methods is necessary. It is time to establish a regular reanalysis of NGS data, every 2-3 years.





## case report: report of a case of diaphragmatic hernia in a 3month infant

پریناز محمود علیلو  $^2$   $^0$  طیبه رضائی  $^1$   $^0$  آیدین محمود علیلو  $^3$ 

دانشگاه علوم پزشکی تبریز، شبکه بهداشت و درمان و مجتمع آموزش سلامت مرند، متخصص طب اورژانس <sup>۱</sup> دانشگاه علوم پزشکی تبریز، شبکه بهداشت و درمان و مجتمع آموزش سلامت مرند، کارشناس ارشد پرستاری داخلی جراحی <sup>۲</sup> دانشگاه علوم پزشکی تبریز، بیمارستان سینا، کارشناس پرستاری <sup>۳</sup>

**Abstract**: Introduction: Congenital diaphragmatic hernia is due to the abnormal growth of the diaphragm during the formation of the fetus and causes one or more abdominal organs to move to the chest and occupy the lung space. As a result, the lungs cannot develop properly. Although this condition is one of the causes of death before birth, timely diagnosis after birth due to respiratory disorder can save the newborn and infant. Case report: A 13month infant, with a history of frequent vomiting after feeding, came to the emergency room with the main complaint of three times of dark brown vomiting. The parents stated that from the beginning of the birth until birth, they had frequent visits to several clinics and hospitals due to frequent vomiting, but the infan's treatment has not been decided. According to the clinical examinations, the decrease in the sound of the lungs was evident in the auscultation of the lungs, and the sound of the lungs the bowel was heard on the left side of the chest. The child's vital signs were in the normal range and slightly tachycardia. In the VBG test, the child had metabolic acidosis. According to these symptoms, a chest scan was performed for the patient and a diaphragmatic hernia was seen in the scan. The infant was sent to an equipped center, diagnosed with diaphragmatic hernia and volvulus and underwent surgery. Conclusion: Failure to timely diagnose diaphragmatic hernia can cause irreparable complications and even cause the death of the infant by causing damage to the lungs. Therefore, in dealing with such cases, detailed clinical examinations, making differential diagnoses and performing scans and graphs are recommended. Accurate ultrasound regarding prenatal anomalies can also help in early diagnosis and treatment Key words: diaphragmatic hernia, infant, disease





## Challenges of home care of cystic fibrosis patients, a thematic analysis

## Leila Taheri <sup>1</sup> <sup>®</sup>, Hakimeh Dehghani <sup>2</sup> <sup>©</sup>, Zahra Abedini <sup>1</sup>, Mohammadreza Modaresi <sup>3</sup>

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Abstract: Background: Cystic fibrosis (CF) is a progressive genetic disorder which involves both internal and external secretion glands. With the increase in the average age of patients with cystic fibrosis, we are faced with a rapid increase in the population of adults suffering from this disease, in whom the complications of cystic fibrosis are becoming increasingly common. This highlights the need for home care and related considerations. Methods: In this qualitative study, we conducted 16 semi-structured interviews with 12 patients with CF and their caregivers. We analyzed the data thematically. Findings: We identified three main themes regarding home care challenges: problems of access to medicine, challenges with airway clearance therapies, and confusion in polypharmacy and diet. The difficult access to vital drugs and the high cost of nebulizers and drugs are one of the important challenges in home care for these patients. Lack of knowledge on how to deal with uncertainties related to drugs, nutrition and activity of these patients is another issues mentioned by the participants which need holistic care plan and a responsive health care system. Conclusion: The past decade has seen remarkable improvements in health outcomes for people with cystic fibrosis and life expectancy for these individuals has increased substantially in our country. Health care providers play an important role in increasing patient and caregivers awareness about CF care. Keywords: Challenges, Home care, Cystic fibrosis, Thematic analysis



