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HEART FAILURE IN CHILDREN - HOW TO OPTIMIZE TREATMENT?

*Simran Jain *Shreepal Jain

Abstract: Heart failure in children represents an important cause of morbidity and mortality in childhood. The recognition of the heart disease in childhood can be challenging as children often have a limited repertoire of presenting signs and symptoms and frequently have illnesses that imitate cardiac disease, such as bronchiolitis, reactive airway diseases and sepsis. Hence, managing heart failure in children requires specific knowledge and skills. There are well-established guidelines for the management of heart failure in the adult population, but the equivalent consensus for paediatric heart failure in children is lacking. This article offers an overview on the etiology, diagnosis, and therapy of pediatric heart failure in children, with a specific focus on practical issues required for management.

Keywords: Heart failure, Cardiomyopathy, Modified ross criteria, Milrinone, Heart transplant.

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Points to Remember

- Heart failure is categorized into 4 stages as per the International Society for Heart and Lung Transplantation (ISHLT) guidelines based on the history, clinical findings and echocardiographic findings.
- Severity grading is based on signs and symptoms and stratified as per modified Ross criteria for <6 years, NYHA for >6 years.
- The diagnosis of HF is an integration of clinical signs, symptoms and relevant investigations which include hematological, echocardiographic and other imaging modalities tailored to individual patient.
- Based on the hemodynamic status, patient should be categorized as either acute decompensated heart failure or chronic compensated heart failure.
- Those with ADHF carry a high risk of morbidity and mortality, thus should be promptly managed in PICU with inotropes, vasodilator and ventilatory support whereas patients with chronic compensated heart failure should get graduated medical therapies based on HF clinical stage.

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MYOCARDITIS IN CHILDREN -APPROACH TO DIAGNOSIS AND MANAGEMENT

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Abstract: Myocarditis is defined as the inflammatory disease of the myocardium, resulting from a wide range of etiologies and they can be diagnosed through a combination of histologic, immunologic and immunohistochemical criteriae. Clinical presentations of myocarditis are often non-specific in nature. The presentation can be subclinical or extensive, manifesting as cardiogenic shock, arrhythmia and fulminant myocarditis leading to rapid progression and sudden death. The diagnosis of myocarditis relies on a range of clinical features and both imaging and non-imaging-based methods. The standard treatment of acute myocarditis in children is not well established and some cases may benefit from immunoglobulin therapy, immunosuppressive therapy or both. The survival rate of children with myocarditis is higher in the childhood population compared to infancy.

Keywords: Myocarditis, Cardiogenic shock, Fulminant myocarditis.

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Points to Remember

- Myocarditis poses significant mortality and morbidity challenges in children due to its diverse presentation and heterogeneous causes.
- Management remains complex, aiming to stabilize vital organs, ensure tissue perfusion and provide adequate ventilation.
- Improved hemodynamic understanding contributes to better survival rates and enhanced treatment approaches.
- Medications contribute to the comprehensive longterm management of myocarditis, addressing both symptomatic relief and improving the underlying cardiac function. The selection of specific drugs depends on the patient's clinical condition and their use should be guided by the treating healthcare provider.
- Regular monitoring and adjustments are essential for optimizing therapeutic benefits.

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PULMONARY HYPERTENSION IN CHILDREN

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Abstract: Pulmonary hypertension in children is a rare disease of diverse etiology. Symptoms of pulmonary hypertension in children are often subtle or misleading and should be suspected in any child with undue shortness of breath, fatiguability or syncope. The diagnostic evaluation of a pediatric patient with pulmonary hypertension is extensive but essential, given the rapid progression of the disease if left undiagnosed and untreated. A targeted approach and an individualized treatment plan for the pediatric patient with pulmonary hypertension is required for better prognosis. Heart and lung transplantation is considered for those who have failed maximal therapy.

Keywords: Pulmonary hypertension, Diagnosis, Targeted approach.

Points to Remember

- Pulmonary hypertension is a rare disease of diverse etiology with a very high morbidity and mortality.
- Thorough etiological evaluation is necessary for all patients with PAH.
- Risk stratification and stepwise therapeutic escalation is the cornerstone for management of PAH.
- Newer treatment modalities like Potts shunt should be considered in patients not responding to medical management.
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RHEUMATIC FEVER AND RHEUMATIC HEART DISEASE: IS IT ON THE DECLINE OR STILL A LINGERING SCOURGE

**Nabeel V Faisal **Saurabh Kumar Gupta

Abstract: Despite rapidly improving socio-economic conditions in India and an apparent decline in the recent past, with a growing population, acute rheumatic fever and rheumatic heart disease continue to remain important public health problems. Any further decline, however, warrants meticulous planning and coordinated efforts. In this brief review, we discuss available data on the burden of acute rheumatic fever/rheumatic heart disease in India.

Keywords: Rheumatic fever, Rheumatic heart disease.

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Points to Remember

- ARF/RHD is on a declining trend globally and in India.
- There is a lack of nationwide data on the exact prevalence of RHD in India.
- Despite an apparent decline, RHD continues to be a public health problem.
- Coordinated efforts are needed for optimal care of patients with ARF/RHD.

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CARDIOMYOPATHIES IN CHILDREN

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Abstract: Cardiomyopathies are disorders of the ventricular myocardium with an estimated incidence of 1 in 100,000 during childhood. The incidence of these disorders peak during infancy and their symptoms are indistinguishable from other common illnesses of infancy including acute respiratory infections and sepsis. The pediatrician requires a high index of suspicion to identify these disorders in a sick child. Hypertrophic cardiomyopathy has a genetic etiology in a majority of cases and appropriate evaluation is important to predict the risk of recurrence in future pregnancies for the parents and also to identify other family members potentially at risk. In this review, we briefly present the etiology, clinical presentation and management of dilated and hypertrophic cardiomyopathy in childhood.

Keywords: Genetic cardiomyopathy, Heart failure, Sudden unexpected death.

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Points to Remember

- Dilated cardiomyopathy is the most common cardiomyopathy in childhood. However, a definitive etiology cannot be identified in approximately half the affected children.
- Structural heart diseases, arrhythmias and nutritional deficiencies are common in children and represent potentially reversible causes of DCM. These should hence be identified and treated early.
- Hypertrophic cardiomyopathy is generally a monogenic disorder and genetic testing is an important part of the evaluation of the child.
- Pediatricians play an important role in the management of children with cardiomyopathy and need to pay particular attention to nutrition, vaccination and growth monitoring.

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BRADYARRHYTHMIAS IN CHILDREN

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Abstract: The common bradyarrhythmia in children include sinus bradycardia and atrioventricular block. Sinus bradycardia is often secondary to underlying noncardiac causes. Congenital complete heart block (CHB) can be isolated or associated with structural heart defects. Isolated CHB is often due to transplacental transfer of maternal autoantibodies. CHB can be diagnosed in-utero by fetal echocardiogram or postnatally by electrocardiogram. It can lead to heart failure or sudden cardiac death. The implantation of pacemaker is recommended for symptomatic patients and for asymptomatic patients with profound bradycardia, ventricular dysfunction, wide QRS interval and prolonged QT interval. This article aims to discuss the etiology, ECG characteristics and approach to management of bradyarrhythmias.

Keywords: Bradycardia, Bradyarrhythmia, Atrioventricular block, Maternal autoantibodies, Pacemaker

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Points to Remember

- Complete heart block can be congenital or acquired.
- Congenital complete heart block is often due to transplacental transfer of autoantibodies in mothers with connective tissue disorders.
- Complete heart block associated with congenital heart defect has a poorer prognosis as compared to those with structurally normal heart.
- Infants with long QT syndrome can present with sinus bradycardia or 2:1 AV block.
- Permanent pacemaker implantation is indicated in symptomatic bradycardia and those with ventricular dilatation and dysfunction.

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GENERAL ARTICLE

ZERO BY 30 - PREVENTING RABIES DEATHS

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Abstract: Rabies is a 100% fatal disease. There is a very high incidence of rabies occurring mainly from dog bites in India. World health bodies have come together to give a call to put an end to deaths from rabies by 2030. This is a preventable disease and the morbidity and mortality can be reduced with timely and appropriate interventions. Both active and passive prophylaxis must be administered along with proper wound toileting. Various vaccine regimens are available as per different world and Indian health bodies. A thorough understanding of the categorisation of bites, available management and need for passive prophylaxis along with active immunisation with vaccines will ensure that there will be zero mortality from rabies in the near future.

Keywords: Rabies vaccine, Rabies immunoglobulins, Monoclonal antibodies, Wound toilet.

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Points to Remember

- Rabies is a slowly progressive, but 100% lethal disease.
- The virus is transmitted from the bites of mammals and wild animals.
- Category III bites require rabies immunoglobulin or monoclonal antibodies to be infiltrated at the site of the bite. This can be administered even if the patient presents late.
- Rabies vaccines are effective when given by the intra-dermal or the intramuscular routes.
- The NCDC guidelines help to guide us to take the correct decisions regarding the category, the need for passive immunisation and the doses of active immunisation that must be given to prevent rabies.

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GENERAL ARTICLE

ENURESIS

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Abstract: Enuresis is one of the most common complaints in pediatric population. It is defined as discrete episodes of urinary incontinence during sleep in children ≥ 5 years of age. It is usually more common in boys and it ceases spontaneously in around 15 percent of affected children every year. Evaluation is based on history and examination, with voiding diary playing a very important role. Enuresis can be treated with urotherapy and committed involvement of the child and parents. Few of them would need medication in consultation with pediatric nephrologist. Desmopressin has found a valuable place in treating enuresis.

Keywords: Enuresis, Evaluation, Children, Voiding diary, Desmopressin Management,

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Points to Remember

- Primary monosymptomatic enuresis is defined by discrete episodes of urinary incontinence during sleep in children above 5 years of age who have never achieved a satisfactory period of night time dryness, with no history of lower urinary tract symptoms or bladder dysfunction.
- Active interventions are warranted as the child gets older, when social pressures increase and self-esteem is affected.
- Management of primary nocturnal enuresis may involve one or a combination of interventions.
- Education and motivational therapies are the initial treatment of choice.
- Enuresis alarms and desmopressin are effective interventions for nocturnal enuresis in children and families who desire active treatment.
- Enuresis treatment, alarms are the most effective long term therapy and have few adverse effects, but requires a long term commitment (usually of three to four months).
- Oral desmopressin works best for children with nocturnal polyuria and normal functional bladder capacity. It is the initial active therapy for children and family.

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DRUG PROFILE

DOSAGE ADJUSTMENTS IN PATIENTS WITH RENAL IMPAIRMENT - ANTIBIOTICS - PART II

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Abstract: Dosage adjustments in patients with renal impairment - antibiotics Part I had highlighted use of some beta lactams, cephalosporins, carbapenems and macrolides in pediatric nephrology. All other commonly used antibiotics and their dosing in children with renal failure, those on peritoneal dialysis (pd), continuous ambulatory peritoneal dialysis (capd), hemodialysis (hd) and continuous renal replacement therapy (CRRT) will be detailed in this article.

Keywords: Renal impairment, dialysis, Continuous renal replacement therapy, Antibiotics.

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Points to Remember

- Aminoglycosides are concentration-dependent killers, interval between doses must be increased in renal impairment; if the renal impairment is severe, the dose itself should be reduced as well.
- Degraded tetracycline (anhydro-4-epitetracycline) may result in renal tubular damage and a Fanconilike syndrome.
- All patients on vancomycin require serumvancomycin measurement (on the second day of treatment, immediately before the next dose if renal function is normal and earlier if renal impairment is present.
- Dosage adjustments in patients with renal insufficiency are recommended for ciprofloxacin, norfloxacin and ofloxacin, but, on the basis of currently available data, adjustments do not appear to be required for the newer drugs like levofloxacin and moxifloxacin.
- In patients with renal impairment: Ethambutol increases the risk of optic neuritis hence preferably avoided in renal impairment.
- No dosage adjustment required for clindamycin, linezolid, chloramphenicol, pyrimethamine.

Note

An attempt to submit some brief guidelines with available data regarding renal dosing of antibiotics was made.

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SURGICAL ARTICLE

GENDER ASSIGNMENT IN DISORDERS OF SEXUAL DEVELOPMENT

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Abstract: This article attempts to clarify the embryological / genetic basis of disorders of sexual developments (DSD)s, to understand the scientific basis for the management of children with DSDs including counselling as regards to gender assignment. DSDs are a group of heterogenous conditions that imposes a heavy psychosocial burden on both the affected child and their family. It is both a social emergency as well as physical and physicalogical one, with conditions that range from ones that are starkly noticed at birth to others that are noticed later in life during the time of sexual maturity. Therefore, it is essential to understand the basis of DSD by understanding embryology of the genitourinary tract as well as the genetic milieuin order to approach these children with DSD with greater sensitivity to offer themadequate help to get an appropriate gender identity.

Keywords: Sexual maturity, Disorders of sexual development, Gender identity.

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Points to Remember

- DSDs are a group of heterogenous conditions that have a heavy psychosocial bearing on both the child and the family ad hence must be managed with utmost sensitivity and care.
- Some rules of thumb are:
 - Testes descend, ovaries do not.
 - Testicular descent is linked to Müllerian ductal regression.
 - Regulation by androgens is exocrine and unilateral.
 - Androgens from non-testicular sources are insufficient to allow development of male genitalia.
 - External virilization is directly proportional to the amount of androgens.
- A phenotypic male child with bilaterally nonpalpable gonads and/or a very proximal and severe hypospadias should undergo investigations for DSDs.
- A phenotypically female child with clitoromegaly, posterior labial fusion and palpable gonads in the labioscrotal or inguinal regions should undergo DSD testing.
- The process of evaluation of DSD starts from a thorough history taking and examination to radiological imaging with USG and MRI, karyotyping, serum studies of hormone levels, genitoscopies and finally diagnostic laparoscopy.

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RADIOLOGY

PLAIN RADIOGRAPHY AND ITS VALUE IN THE ASSESSMENT OF PEDIATRIC CARDIAC PATIENTS - PART I

*Sudeep Verma

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CASE REPORT

A CHAOS TO CONSIDER

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Abstract: Congenital high airway obstruction syndrome, a rare entity is a complete or partial obstruction (atresia/stenosis) of the upper airways (laryngeal/tracheal) in neonates and was first described by Hedrick in the twentieth century. Despite modern imaging advancements, it is still being identified only postpartum and has poor outcomes without appropriate early interventions. We report one such case from our institution.

Keywords: Congenital high airway obstruction syndrome, EXIT procedure, Laryngeal atresia.

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