Lest we forget - Experiences in Heart Failure during Infancy and Early Childhood

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Foreword from the Editor:
Timely identification and management of congenital heart diseases and other cardiac problems in infants is tantamount to preventing death, inoperability and long-term sequel. This also helps to anticipate and treat heart failure and tackle issues which could precipitate or worsen this complication till a corrective surgery could be carried out. It is important to educate the parents as well as primary health-care providers who are directly involved in management of such children. Advances in technology have facilitated diagnosis of critical congenital heart problems as early as pre-natal stage for certain diseases and institute safe confinement at centres where sick new-borns could either be operated upon or stabilized and referred for surgical or interventional procedures. However, it is easy to overlook diagnoses of cardiac problems in infancy and early childhood when working in busy conditions or resource-poor primary care settings. The author narrates some of the clinical cues which would help a pediatrician to detect and manage such conditions in this population, providing a series of real-life experiences.

Diagnostic dilemmas in pediatric heart failure

Starting off with the era when echocardiographic evaluations were not easily available; asymptomatic infants with holosystolic murmur without cardiomegaly were clinically diagnosed to have small ventricular septal defects (VSD) after ECG and x-ray. On follow up, majority of such children would gain weight normally and continue to be asymptomatic. However a few of those children presented later with cardiomegaly and heart failure. Then only we became aware of the entity of sub-pulmonic location of VSD and risk of development of aortic regurgitation. We realized that delineating the anatomical location of a VSD is important in deciding management and follow up. In the present practice such instances are uncommon since such infants are subject to echocardiographic evaluation and advised surgery appropriately.

However, echocardiography has to be in conjunction with a good clinical assessment incorporating x-ray and ECG. A few conditions may be easily overlooked otherwise, such as anomalous pulmonary venous drainage, atrial septal defects (ASD) of the sinus venosus type, coronary artery anomalies, etc. Some of the conditions might not present in the early neonatal period due to slow decline of the elevated pulmonary vascular resistance. A follow up assessment of new born baby without any identifiable abnormalities will help to uncover such issues.

Case 1: A seven-day old baby delivered at term, vaginal delivery with adequate birth weight was normal for first few days suddenly developed respiratory distress. The baby was relatively quiet when brought in and saturation was normal with mild tachypnea. All peripheral pulses were palpable, with good femoral artery pulsations. Respiratory system examination was within normal limits. X Ray showed clear lungs, normal vascularity and minimal cardiomegaly. ECG showed sinus rhythm, tachycardia, right ventricular hypertrophy and QRS axis of +105 degrees. Echocardiography was reported normal. Arterial blood gas analysis was normal. Baby was followed up, the symptoms improved over the next few days. Suddenly baby became severely distressed and irritable. Repeat clinical examination showed gross cardiomegaly, and the femoral pulses were hard to palpate in the struggling baby. A repeat echocardiography revealed coarctation of aorta. Pediatricians and pediatric cardiologists have experienced similar dilemma in diagnosing neonatal aortic coarctation in babies who become sick.

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intermittently. A clinical examination when the baby is sick and irritable might not permit assessment of the lower limb pulses diligently when the pulsations might be truly impaired. When such babies become more stable as the ductus opens up, femoral artery pulsations might be passed off as normal. A carefully repeated clinical examination, assessment of lower limb pulses and saturation and repeated echocardiographic evaluation might help to diagnose aortic coarctation which will be life-saving in such neonates.¹

Case 2: In rare occasions, babies presenting with failure to thrive can continue to baffle the pediatricians. A six month old baby with failure to thrive was referred to the unit since no cause could be identified even after extensive evaluations including echocardiogram. The baby was tachypneic at rest and apart from being underweight, and having a larger than normal sized head and mild cardiomegaly, nothing else could be detected. The baby was being worked up for renal tubular acidosis and organic acidiemias. During a detailed anthropometric evaluation, the Resident Doctor could elicit a bruit over the head. A neurosonogram through an open fontanelle and subsequent MRI confirmed the diagnosis of vein of Galen malformation. Since then we made it a point, to measure head circumference and auscultate the head in children with Congestive Heart Failure (CHF), especially when the cause was not obvious which helped in a few subsequent occasions.

Case 3: In another example, a 3.2 kg baby born at term by vaginal delivery had an uneventful neonatal period with expected weight-gain; however the mother noted that the baby had rapid breathing with suck rest cycles, which gradually progressed over next few days. When he was brought at 56 days his weight-gain was inadequate, normal development with a good smile on his face. Clinical examination showed he was in failure, with a pulse of 170 rate and normal volume, cardiac apex was in fourth space outside mid-clavicular line, heart sounds were soft with a prominent S3, faintly audible systolic murmur at apex, though there was no mid-diastolic murmur. A working diagnosis of moderate VSD was made as presentation was classical. X Ray showed cardiomegaly, but without increased pulmonary vascularity. ECG revealed significant Q waves in leads I and aVL. The echocardiogram revealed the diagnosis of anomalous origin of left coronary artery from pulmonary artery (ALCAPA). A diagnosis of ALCAPA should be entertained in babies just about one month old, presenting with unexplained excessive cry and features of CHF of recent onset.

Case 4: This is a story of a baby born at term with history of birth asphyxia, discharged after two weeks of NICU stay. The baby was found to be hypotonic from the outset which was attributed to birth asphyxia. Follow up at six weeks revealed established social smile and usual alertness, but the baby was hypotonic with absent deep tendon reflexes. Diagnosis at this stage was revised as motor unit disorder spinal muscular atrophy/congenital myopathies. Basic level investigations like Creatinine Phosphokinase (CPK) were ordered and we were planning for nerve conduction studies and genetic analysis. Repeat clinical examination showed soft hepatomegaly, and the baby appeared dyspnoeic, which might not have been noticed initially because of any appreciable chest in-drawing or grunting due to the muscle weakness. This prompted a careful cardiovascular re-examination, which revealed cardiomegaly and led on subsequently to the diagnosis of Pompe’s disease. This case highlights that a sequential pattern of history, detailed systemic examination and investigations help to uncover the diagnosis when one is at cross-roads. That case was proved to be a case of Pompe’s disease. And as we can expect, macroGLOSSIA was the last feature to be examined for. The lesson I learnt was that in such babies, clinical features of CHF are modified due to the lack of muscle power. In hypotonic babies with muscle disorders due to mitochondrial and metabolic problems always assess the heart.

Case 5: Long back in seventies, a four weeks old male baby with normal birth weight was brought for evaluation of poor weight-gain. The baby was emaciated but alert and active, with tachycardia, cardiomegaly, normal heart sounds and no murmurs. Baby had prominent eyes which permitted us to look at the mother’s face. She had classical eye signs of hyperthyroidism. The baby’s thyroid profile showed high thyroxine level with suppressed TSH.

Management considerations in CHF during infancy and in early childhood

Till the turn of the present century, pediatricians in Kerala used to encounter older children with HF, consequent to rheumatic valvular heart diseases including acute rheumatic fever and congenital heart diseases, remaining uncorrected for various reasons. Presently major share of heart failure in children in the state are due to congenital heart diseases who present during infancy and early childhood itself. The definitive management is appropriately timed surgery or intervention as the case may be. However, it is important to be aware of ancillary aspects of care of such patients.

(a) Nutrition

A study from Kerala has previously reported high prevalence of malnutrition in children with various congenital heart diseases undergoing corrective intervention.² Infants with heart failure need more energy during the resting stage due to extra work of breathing, myocardial energy requirements,
thermoregulation. Moreover, the percentage of lean body mass is higher with lesser fat stores, which also increases the basal metabolic rate\(^6\). The energy requirement during physical activity can go up by as much as 250 % of age-matched healthy infants\(^3\). Milk & milk products from non-human sources have higher sodium content and could pose problems in HF and breast milk appears to be the best source of nutrition in term infants. The energy required for sucking can be done away with in sick infants by appropriately maintained oro-gastric tubes at home or in hospital with adequate training\(^6\). To promote adequate milk secretion, the mother should be reminded to put the baby to the breast periodically without exhausting the baby. If the breast milk quantity and/or weight-gain seem inadequate, the mother can be instructed to add a few drops of coconut oil to the breast milk. Human milk fortifiers, formula feeds and micronutrient supplementation are generally not required in term normal birth weight infants on breast milk. Complementary feeds need be initiated at 6 months in an infant born at term judiciously.

**b) Drugs**

Excellent resources are available for drug therapy of cardiac diseases in children including a consensus statement from the Working Group on Management of Congenital Heart Diseases in India\(^6\). Digoxin is still one of the mainstay drugs prescribed in pediatric HF and caution regarding correct dose has to be exercised considering factors like term vs pre-term, weight, presence of myocarditis, renal dysfunction, etc. Digoxin is available as an elixir in the concentration, 50μg/ml, and the daily maintenance dose is 10μg/kg/day for a term-born baby. For a 6 kg baby, it will be 60μg per day, which can be remembered easily as 0.6ml twice daily, 0.5ml twice daily for 5kg child and so on; i.e. [weight (kg) / 10] mL two times daily. Potassium supplementation at 1-1.5mEq/kg/day should be undertaken with diuretic therapy, especially if the dose of Furosemide is higher than 2mg/kg/day, unless there is concomitant use of renin-angiotensin-aldosterone system (RAAS) blockers. Vasodilators like Angiotensin converting enzyme inhibitors (ACEI) are prescribed in presence of large left to right shunts and hyperkinetic pulmonary hypertension in the absence of systemic hypotension. They should not be used in children with obstructive lesions. Occasionally we face situations when the dosage of medications is not adequate, resulting in less control of symptoms.

During follow up, the dose has to be titrated according to the weight of the baby. For some medications like beta blockers and ACEI, tablet preparations meant for use in adult patients have to be broken down which also can potentially result in wrong dosage.

**c) Other issues**

Universal vaccination should be recommended as in a healthy infant. Pneumococcal conjugate vaccines are especially recommended in infancy. Bronchiolitis is a particular threat in these infants and result in fatalities. Monthly injections of Palivizumab are recommended against respiratory syncytial virus (RSV) infections in these children in many countries during the RSV season. Prevention or management of anaemia and deworming should be carried out appropriately. Monitoring of development is important in these children as motor mile-stones could get delayed. Genetic evaluation and psychosocial counselling, counselling about modified history and follow up are other important measures.

**Conclusions**

Heart failure in newborn and infancy can occasionally present with diagnostic challenges. Prompt identification of the underlying cause and definitive treatment is of utmost importance. Periodic reassessment helps to minimize overlooking the diagnosis. A good supportive care helps in achieving optimal surgical outcomes.

**References**