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Review Article

Emergency Management of Critical CHD in Limited Resource Settings

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ABSTRACT

Critical congenital heart disease (CCHD) is defined as lesions that are ductal dependant and/or require surgical or catheter intervention in neonatal period. The incidence of congenital heart disease is estimated to be 8-10 per 1000 live births and one third to half of these CHD are critical CHD, requiring urgent intervention. Considering high number of births in India, the need to diagnose and manage such newborns gains importance. Newborn with CCHD are apparently well at birth. Thorough Physical examination at birth does not provide any additional helpful information, as many normal newborns at birth may have murmurs, and many critical CHD may not have any murmur. Not many hospitals in developing countries like India have access to pediatric cardio surgeon. The article reviews management and issues in children with critical CHD in limited resource settings.

Keywords: critical CHD, ductal dependent heart disease, prostaglandin, pulse oximetry screening.

INTRODUCTION

Critical congenital heart disease (CCHD) is defined as lesions that are ductal dependant and/or require surgical or catheter intervention in neonatal period. [1] The incidence of congenital heart disease is estimated to be 8-10 per 1000 live births and one third to half of these CHD are critical CHD, requiring urgent intervention. [2-4] No data exists about incidence of critical CHD from India as many children are still delivered at whom do not have ready access to healthcare facilities. However, considering high birth rate in our country, number of children with critical CHD might actually be huge.

Newborn with CCHD are apparently well at birth. Thorough Physical examination at birth does not provide any additional helpful information, as many normal newborns at birth may have murmurs, and many critical CHD may not have any murmur. Ainsworth et al [5] studied 7204 newborn that first underwent physical examination for evaluation of cardiac disease by senior house officers, and then subsequent echo was done. Murmurs were detected in 46 neonates, out of whom only 24 had CHD. However, another 32 infants with normal examination were found to have CHD on echo. [5]

As neonates with critical heart lesions are asymptomatic at birth, they are likely to be discharged from hospital without being diagnosed and in lack of access to health facilities, many of them die without being diagnosed. [6] Even in developed countries, around a quarter of children with CCHD were discharged from hospital without diagnosis. [7] Many children in India are still born at home, and those who are born at hospital, due to overcrowding and limited resources are discharged from hospital early. Since there is no program for neonatal screening in India, and lack of any signs and symptoms

in first few days of life, these children are discharged from hospital undiagnosed.

The basic difference between fetal circulation and postnatal circulation is place of oxygenation. While placenta is responsible for oxygenation in fetus, postnatally, lungs perform the function of oxygenation. There are various shunts in fetus (ductus arteriosus, ductus venosus, and foramen ovale) which close after birth. Because of these shunts, even severe cardiac malformations maintain appropriate circulation in utero, but the child starts collapsing postnatally when the ductus arteriosus close. Functional closure of the ductus arteriosus occurs within 10 to 15 hours after birth by constriction of the medial smooth muscle while anatomic closure is completed by 2 to 3 weeks of age. Anatomic closure of ductus arteriosus occurs due to permanent changes in the endothelium and subintimal layers of the ductus. This is the period when critical cardiac lesions present. The presentation may be delayed as hypoxia and acidosis causes relaxation of ductus arteriosus. [8,9]

Patients with CCHD will have a severely compromised systemic or pulmonary circulation and cannot tolerate the transition from fetal circulation to postnatal circulation after birth. These children depend on the central shunts, especially patent ductus arteriosus (PDA) for adequate circulation. These lesions which are dependent on blood flow through the PDA for adequate circulation are collectively referred as "ductal-dependent lesions". Ductal dependent lesions can be classified as ductal dependent systemic circulation (hypoplastic left heart syndrome, interrupted aortic arch, critical coarctation of aorta, TAPVC) or ductal dependent pulmonary circulation (tetralogy of fallot with pulmonary atresia, critical PS, tricuspid atresia, ebstien's anomaly,) and ductal dependant mixing lesion (TGA with intact septum), where duct is required to maintain mixing between systemic and pulmonary circulation. Patients with ductal dependent lesions will present with severe cyanosis, shock or collapse when PDA constricts within hours or days after birth.

Critical CHD presents wither cyanosis, respiratory distress or shock. Neonates with CCHD presenting in ER may be a diagnostic challenge, as differential diagnosis includes sepsis, inborn error of metabolism, critical heart disease and cardiac arrhythmias, endocrine disorders, trauma and poisoning. Presentation of CCHD can with cyanosis, shock or both. While evaluating a neonate with cyanosis, respiratory cause of cyanosis can be differentiated by cardiac cause by hyperoxia test, where response to 100% oxygen inhalation is tested on arterial pO_2. If pO_2 fails to rise more than 150 mm Hg on 100% oxygen inhalation, or rise in pO_2 is not more than 10-30 mm Hg, then it indicates cardiac cause of cyanosis. Common CCHD presenting with cyanosis includes TOF with severe PS, TGA, Tricuspid atresia, TAPVC and Truncus arteriosus. CCHD presenting with shock includes critical outflow obstruction like critical AS, Hypoplastic left heart syndrome, critical coarctation and interrupted aortic arch. They are more frequently missed as most of them are treated as septic shock. Examination shows discrepancies in pulses and blood pressure of upper and lower limbs.

Initial management of both group of CCHD is under same lines. There may be technical reasons for delay in confirming diagnosis due to non availability of echo round the clock at many centres. But this should not delay the management. The aim of medical intervention is to restore fetal circulation/interrupt transition to neonatal circulation. These children are volume deficient due to poor feeding and vomiting. Bolus of normal saline in aliquots of 10 ml per kg should be given initially, keeping a check on liver size. Volume expansion helps to restore perfusion, improve acidosis and also assists opening of ductus arteriosus. NSAID including oral paracetamol causes closure of ductus arteriosus by inhibiting prostaglandin synthesis, [10] and in any suspected or undiagnosed child presenting

with fever/shock/differential cyanosis, any NSAID including paracetamol should be avoided.

Alprostadil infusion (PGE1) should be started as soon as diagnosis is made or even suspected, which helps in keeping ductus open, or reopens closed ductus. It is administered in dose of 0.05 to 0.1 µgm/kg/min. Complications of alprostadil includes apnea, flushing and hypovolemia. Children who are given prostaglandins may need mechanical ventilation, and if they are to be shifted to higher centre on prostaglandin infusion, they should be electively ventilated. Oral prostaglandin (PGE2/dinoprostone) is as effective as alprostadil in opening of ductus, and has less chances of apnea. [11,12] Hence in limited resource settings or non availability of persons trained in intubation, oral prostaglandin PGE2 Dinoprostone can be given in doses of 25 µgm /kg. However, not many reports are available of its use from India, and experience about its use is limited. It is to be kept in mind that in obstructive type of TAPVC, prostaglandin infusion is not effective, and ECMO is only available method of stabilization. Prostaglandins may reopen ductus in neonate upto 30 days of age. Accompanying acidosis aggravates pulmonary vasoconstriction, and hence correction of acidosis by administering bicarbonate is recommended. Inotropes are required to maintain adequate tissue perfusion. Only after child is stabilized with these interventions, the child should be referred to pediatric cardiologist for definitive surgical interventions.

Neonatal screening for CCHD was started in US in 2011. This involves pulse oximetry of newborn, at 24 to 48 hours of age, just before discharge. Screen is considered positive if saturation in Right upper limb is less than 95% or difference in saturation between preductal and postductal limb is more than 3%. All those who are screened positive are subsequently sent for echo. However, the common cause of difference in saturation of more than 3 % between two limbs is persistent pulmonary hypertension. The normal saturation values in children living at higher altitudes were found to be somewhat lower. [13]

Though universal pulse oximetry screening is well known to reduce mortality, even in developing countries, [14] there are still many barriers in implementing it. First and foremost, pediatric cardiac surgeons are available only in few centers in India, some of them in private sector and available at a cost not all can afford. Even government medical colleges in India lack pediatric cardio surgeon. Thus, biggest hurdle in developing countries is the availability and cost of treatment even when appropriately diagnosed.

The disease should be diagnosed early as timely intervention significantly reduces mortality. [15] Antenatal diagnosis is not possible in majority. Freidberg et al identified infants with major CHD in 3 major centers in California, and found that only 28% of children were antenatally diagnosed with TAPVC, TGA and left sided obstructive lesions had lowest prenatal detection. [16] With poor antenatal care in developing countries, detection of antenatal CHD is even lower, and in absence of universal neonatal screening, more neonates are bound to be discharged from hospital undiagnosed, presenting later with cyanosis/shock.

Neurodevelopmental outcome of child with CCHD is also a concern. Newly acquired brain injuries are commonly detected radio logically in term newborn with CHD, [17-20] which consist of white matter injuries similar in pattern to disease of premature newborns. These newly acquired brain injuries are supposed to be due to compromised cerebral perfusion during cardiopulmonary bypass. However, increasingly body of literature suggests that these children preoperative neurological injuries, which is contributed by intrauterine hemodynamic alterations, congenital brain anomalies and acquired brain injury related to prolonged cyanosis or hypo perfusion after birth. [21] The associated congenital

anomalies with critical CHD should also be searched for and informed to parents.

Considering the vast terrain of India, making a universal cutoff of SpO_2 is not feasible as it has been shown to vary with altitude, with lower SpO_2 at higher altitudes. At present there is no ongoing neonatal screening programme. Neonatal screening for CCHD is still due in India, and should be started, at least in tertiary care level. Along with this, efforts should be made to provide more pediatric cardio surgeons available in government sector, to reduce burden of cost to the needy, unaffordable group of children.

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