

Theme: Pediatric Cardiology

Indian Guidelines for indication and timing of intervention in congenital heart diseases (*Ann Pediatr Cardiol.* 2019;12:25486)

Various international guidelines on management of congenital heart disease (CHD) are available, but their applicability to Indian population is likely to be limited, as majority are not diagnosed in antenatal period and often present late in the course of disease. Further complications arise due to underweight, malnutrition, and comorbidities such as recurrent infections and anemia. Also, the late presenters have advanced level of pulmonary hypertension, ventricular dysfunction, hypoxia and polycythemia, which lead to suboptimal outcomes and long periods of hospital stay. In this document, evidence-based guidelines for Indian scenario are presented, which include (i) indications and optimal timing of intervention in common CHD; (ii) followup protocols for patients who have undergone cardiac surgery/catheter interventions for CHD; and (iii) indications for use of pacemakers in children.

Differences in cardiovascular manifestation of Marfan syndrome between children and adults (*Pediatr Cardiol.* 2019;40:393-403)

Cardiovascular findings associated with Marfan syndrome include aortic aneurysm and aortic dissection or rupture, aortic regurgitation, mitral valve prolapse and regurgitation, tricuspid valve prolapse and regurgitation, pulmonary artery dilatation and primary cardiomyopathy. The authors aimed to evaluate cardiovascular system in 44 children and 57 adults with Marfan syndrome, and describe the type, incidence and severity of these findings. The most common cardiovascular abnormality was aortic root dilatation (81.2% of patients). They observed that both adults and children had similar high rates of aortic root dilatation ($P=0.20$). Similarly, there was no significant difference with regard to the prevalence of aortic valve regurgitation ($P=0.49$) and mitral valve prolapse ($P=0.69$) among children and adults. These findings indicate that the aforementioned abnormalities develop in early childhood, and therefore, these may be used in the early identification of patients with Marfan syndrome. Other assessed abnormalities, which included mitral valve regurgitation ($P=0.002$), pulmonary artery dilation ($P=0.025$), aneurysms of aortic arch ($P=0.04$), descending thoracic aorta and abdominal aorta ($P=0.015$) were found mostly in adults, and thus, are of less use in the early detection of Marfan syndrome.

Antenatal therapy for fetal supraventricular tachyarrhythmias (*J Am Coll Cardiol.* 2019;74:874-85)

As standardized treatment of fetal tachyarrhythmia is not established, this multi-centric study aimed to evaluate the safety and efficacy of protocol-defined transplacental treatment for fetal supraventricular tachycardia (SVT) and atrial flutter (AFL). In 49 singleton pregnancies, from 22 to <37 weeks of gestation, with sustained fetal SVT or AFL ≥ 180 beats/min, transplacental treatment using digoxin, sotalol, and flecainide was administered. The primary endpoint was resolution of fetal tachyarrhythmia.

Secondary endpoints were fetal death, preterm birth, and neonatal arrhythmia. Fetal tachyarrhythmia resolved in 89.8% (44 of 49) of cases overall, and in three out of four cases of fetal hydrops. Preterm births occurred in 20.4% (10 of 49) of patients. Maternal adverse effects were observed in 78% (39 of 50) patients. Serious adverse events occurred in one mother and four fetuses, thus resulting in discontinuation of protocol treatment in four patients. Two fetal deaths occurred, mainly caused by heart failure. Neonatal tachyarrhythmia was observed in 31.9% (15 of 47) of neonates within 2 weeks after birth. Protocol-defined transplacental treatment for fetal SVT and AFL was effective and tolerable in 90% of patients. However, serious adverse events may occur in fetuses, and that tachyarrhythmias may recur within the first 2 weeks after birth.

Ventricular-arterial coupling: A novel echocardiographic risk factor for dilated cardiomyopathy (*Pediatric Cardiol.* 2019;40:330-8)

Ventricular-arterial coupling pertains to energy losses in overcoming resistance during energy transfer from ventricle to systemic arterial circulation, for maintenance of organ perfusion. It is measured by the ratio of arterial elastance (Ea) to left ventricular (LV) end-systolic elastance (Ees) ($VA \text{ coupling} = Ea/Ees$). Echocardiographic LV elastance (Ees) is calculated as $(0.9 \times \text{systolic blood pressure}) \div (2D \text{ echocardiographic end-systolic volume})$. Echocardiographic arterial elastance (Ea) is calculated as, $(0.9 \times \text{systolic blood pressure}) \div (2-D \text{ echocardiographic stroke volume})$. The authors sought to determine if VA coupling is different in pediatric patients with dilated cardiomyopathy (DCM) compared to normal controls, and to determine if VA coupling is different in pediatric DCM with poor outcome vs those without. Authors analyzed data from 48 patients and 97 age- and gender-matched controls, studied their outcomes at 2 years after entry to the cohort. Twenty-seven (56%) patients reached composite endpoint (mechanical circulatory support, transplant, or death) by the end of 2 years. Patients with DCM had significantly higher heart rate, lower blood pressure, higher LV dimensions, lower EF, and poor tissue Doppler indices, compared to normal controls. Ventricular elastance was significantly lower in DCM group and arterial elastance and VA coupling were significantly higher in DCM group ($P<0.001$). The comparison of the two DCM groups (with poor outcome and good outcome) revealed that patients with poor outcome were significantly younger, had acute heart failure, had worse NYHF/Ross class, and higher heart rates, higher arterial elastance, lower ventricular elastance and higher VA coupling compared to the patients with good outcome. CART risk stratification revealed that among all the variables, VA coupling ratio ≥ 2 , was the top discriminator of poor outcome with no additional variables needed to stratify. Kaplan–Meier curve analysis demonstrated patients with VA coupling ratio ≥ 2 had significantly poor event-free survival ($P=0.001$). Hence pharmacotherapy targeting these variables may improve prognosis and outcomes.

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