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Leading Article

Intervention in Congenital and Structural Heart Diseases in Children

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Catheter-based techniques, whether palliative or corrective, are the accepted therapy for many congenital and structural cardiac defects. In the past 2 to 3 decades, the field of pediatric interventional cardiology has experienced significant growth. Interventional therapy has become an acceptable alternative treatment for many CHD, including closure of atrial defects, muscular ventricular septal defects (VSDs), patent ductus arteriosus (PDA), dilation of stenotic valves (aortic and pulmonary), and dilation of stenotic vessels (branch pulmonary arteries, coarctation of the aorta COA).

Neonatal cardiac lesions and situations requiring immediate intervention includes TGA (Transposition of Great Arteries) associated with restrictive atrial septum, HLHS (Hypoplastic Left Heart Syndrome) or critical Aortic Stenosis, PDA dependent pulmonary or systemic circulation. Interventions done for these lesions are urgent Balloon atrial septostomy, Urgent balloon valvuloplasty, PDA stenting or BT shunt respectively.

At present, lesions with the greatest potential benefit from fetal interventions are fetal arrhythmias, severe aortic or pulmonary stenosis, and HLHS.

Atrial level shunts, such as atrial septal defect (ASD) and patent foramen ovale, are common congenital cardiac defects. Patients with ASDs are usually asymptomatic in the first 2 decades of life. Usually, if the Qp/Qs ratio is more than 1.5:1, patients experience symptoms of shortness of breath and fatigue. In 1974, King and Mills performed the first successful transcatheter closure of a secundum ASD.

Ventricular septal defects are considered the most common cardiac abnormalities found in children, accounting for approximately 30% of all defects. Large VSDs are unlikely to close spontaneously, and patients with such defects will present with signs and symptoms of congestive heart failure and failure to thrive. Currently PM VSD and Muscular VSD's are closed interventionally by Device. Device closure of ventricular septal defect remains a challenge, as the majority of VSD's exist in anatomically unfavorable locations within the ventricular septum.

Patent ductus arteriosus represents 5% to 10% of all CHD, excluding those in premature infants. In infants beyond the neonatal period, device and or coil closure can be done. Aortopulmonary windows are rare forms of CHD. Large defects are usually closed surgically. If the defect is small, percutaneous device closure may be feasible.

Isolated valvular pulmonary stenosis (PS) represents 80% to 90% of all patients with right ventricular outflow tract obstruction, and 8% to 10% of all patients with CHD. Indications for Balloon pulmonary valvuloplasty include symptomatic patients, asymptomatic patients with severe PS (gradient 80 mm Hg), or evidence of right ventricular hypertrophy. Variable modalities of percutaneous treatment regimens include balloon pulmonary angioplasty, cutting balloons, and intravascular stent placement. Stenting of branch pulmonary arteries is frequently used in children with pulmonary artery stenosis and/or hypoplasia.

In selected infants with membranous pulmonary atresia and adequately sized right ventricle, perforation of the membrane followed by balloon dilation has been successful in creating an open right ventricular outflow tract.

Left-Sided Obstructive Lesions like Valvular Aortic valve Stenosis (AS) occurs in approximately 3% to 6% of patients with CHD. Indications for intervention include neonates with critical AS, irrespective of the gradient and infants and children with peak-to-peak gradient across the valve of more than 55 mmHg.

Coarctation of the aorta accounts for 5% to 8% of all CHD. Indications for balloon dilation of COA include

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hypertension proximal to the coarctation with a resting systolic pressure gradient across the narrowed segment 20 mm Hg or angiographically severe coarctation with extensive collaterals.

Endovascular stent implantation has become an accepted modality for treatment of native and recurrent coarctation of the aorta in children and adults. Self expandable stents can be deployed in younger children >25 kg body weight. Congenital mitral valve stenosis is a rare lesion. Balloon angioplasty is a well-known treatment modality for treatment of rheumatic mitral valve stenosis.

After fenestrated Fontane operation, sometimes a significant right-to-left shunt through fenestrations, causing systemic hypoxemia and desaturation. These fenestrations are often amenable to closure in the cardiac catheter laboratory, using the same type of devices that would be used to close an ASD.

Aortopulmonary Collateral vessels occur in a wide variety of complex cyanotic heart diseases and. They may be arterial or venous and may shunt left to right or right to left. As they are often difficult for the surgeon to access, they may be occluded with coils or occluder devices in the cardiac catheter laboratory.

Extracardiac conduits may be used to connect the sub-pulmonary ventricle to the pulmonary arteries in patients with complex congenital disease. These conduits can develop obstruction secondary to severe angulation, calcification, sternal compression, or tissue proliferation, mainly at anastomotic sites. Balloon dilatation of stenosed conduit causes relief of symptoms. Stent implantation has demonstrated better long-term outcome, yet with recognized risks

Tetralogy of Fallot accounts for 3.5% of infants born with CHD. Pulmonary regurgitation, residual peripheral arterial pulmonic stenosis, and small residual VSDs are common after surgical repair of tetralogy of Fallot. The use of balloon angioplasty and the cutting balloon has been successful in releasing residual peripheral arterial pulmonic stenoses that are usually more distal than the surgeons can reach. In situations where a significant residual defect is noticed, percutaneous device closure may take place if feasible technically.

Pulmonary regurgitation is common following repair of tetralogy of Fallot. Although well tolerated for some years, Pulmonary valve replacement is the treatment of choice when indicated.

The spectrum of transcatheter procedures available for the treatment of children and adults with congenital heart disease has rapidly increased over the last three decades. While the technical skills of the operator combined with a sound anatomic and hemodynamic understanding of a patient's condition remain without a doubt, the most important ingredients for successful outcome of an interventional catheter procedure, the choice of the appropriate equipment is almost equally as important for a successful outcome. For any procedures to be performed at any particular institution, minimal specific skills are required, special techniques must be mastered and maintained, and a large inventory of specialized and expensive catheters and devices must be stocked to offer the patient an optimal procedure.

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