Symposium on Pediatric Cardiology-I

Diagnosis and Management of Acyanotic Heart Disease: Part I - Obstructive Lesions

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Abstract. In this review, the clinical features and management of most commonly encountered acyanotic obstructive cardiac lesions are discussed. Mild lesions, especially in children are usually asymptomatic while neonates and infants may present with symptoms. Ejection systolic murmurs in patients with pulmonic and aortic stenosis and decreased femoral pulses and blood pressure difference (>20 mmHg) between arms and leg in patients with aortic coarctation are usually seen. Clinical diagnosis is not difficult and the diagnosis can be confirmed and quantitated by non-invasive echocardiographic studies. Whereas surgical intervention was used in the past, balloon dilatation appears to be effective in the treatment of these lesions.

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Congenital heart defects (CHDs) may be classified into acyanotic and cyanotic, depending upon whether the patients clinically exhibit cyanosis. The acyanotic defects may further be subdivided into obstructive lesions and left-to-right shunt lesions. The cyanotic defects, by definition, have right-to-left shunt. In this review obstructive cardiac lesions will be discussed. The objective of this review is to describe the important findings in history, physical examination and laboratory studies that are suggestive of the diagnosis of the respective obstructive lesions and to discuss the available options in the management of these defects.

OBSTRUCTIVE LESIONS

When there is a significant narrowing of a valve or a blood vessel, there is a higher pressure proximal to the obstruction compared to the distal pressure; this pressure gradient is necessary to maintain flow across the stenotic site. Hypertrophy of the cardiac chamber proximal to the obstruction and flow disturbance across the site of obstruction and their effects will determine the clinical features. More commonly encountered obstructive lesions, namely pulmonary stenosis, aortic stenosis and coarctation of the aorta will be reviewed.

Pulmonary Stenosis

The obstruction can be at valvar, subvalvar or supravalvar sites or in the branch pulmonary arteries. Valvar stenosis is the most common type and will be discussed in this section. Valvar pulmonary stenosis (PS) constitutes 7.5% to 9.0% of all CHDs. The pathologic features of valvar stenosis vary, but the most commonly found pathology is what is described as “dome shaped” pulmonary valve with fusion of the thickened pulmonary valve leaflets. Hypertrophy of the right ventricle (proportional to the degree of obstruction) and dilatation of main pulmonary artery (not related to the severity of obstruction) are also seen.

Symptoms: Children with PS usually present with asymptomatic murmurs, although they can present with signs of systemic venous congestion (usually interpreted as congestive heart failure) due to severe right ventricular dysfunction or cyanosis because of right-to-left shunt across the atrial septum.

Physical Findings: The right ventricular and the right ventricular outflow tract impulses are increased and a heave may be felt at the left lower and upper sternal border. A thrill may be felt at the left upper sternal border and/or in the suprasternal notch. The first heart sound may be normal or loud. The second heart sound is variable, depending upon the degree of obstruction and will be detailed latter in this section. An ejection systolic click is heard in most cases of valvar stenosis. The click is heard best at the left lower, mid and upper sternal borders and varies with respiration (decreases or disappears with inspiration). An ejection systolic murmur (Figure 1) is heard best at the left upper sternal border and it radiates into infraclavicular regions, axillae and back. The intensity of the murmur may vary between grades II to V/VI; the intensity is not necessarily related to the severity of the stenosis.
Clinical assessment of severity. The timing of the click, the extent of splitting of the second sound, the intensity of the pulmonary component of the second sound, the length (duration) of the murmur, and timing of peaking of the systolic murmur are usually suggestive of the severity of pulmonary valve obstruction (Fig 1). In mild cases of pulmonary valve narrowing, the click is clearly separated from the first heart sound, almost normal splitting of the second heart sound with normal or slightly increased pulmonary component of the second sound is heard, and an ejection systolic, diamond-shaped murmur that peaks early in systole and ends way before the aortic closure of the second heart sound is appreciated. The findings in moderate PS include an ejection systolic click that is much closer to the first heart sound than in milder forms (Fig 1), widely split second sound with diminished pulmonary component of the second sound and an ejection systolic murmur that peaks in mid to late systole and ends just before the aortic component of the second sound. The features of severe valvar PS are an ejection systolic click which is either not present or falls so close to the first heart sound that it becomes inseparable from it, markedly increased splitting with a soft or inaudible pulmonary component of the second heart sound, and a long ejection systolic murmur that peaks late in systole and extends beyond the aortic component of the second sound so that the latter cannot be heard. The loudness of the ejection systolic murmur does not indicate the severity of obstruction but rather its duration and time of peaking; the longer the murmur and the later it peaks, the more severe is the PS. Similarly, the shorter the time interval between the first heart sound and ejection click, the wider the splitting of the second heart sound, and softer the pulmonary component, the more severe is the degree of pulmonary valve obstruction.

Noninvasive evaluation: Chest X-ray, in most cases, shows no cardiomegaly, but a characteristically dilated main pulmonary artery segment (post-stenotic dilatation) is visualized (Fig. 2). The magnitude of pulmonary artery dilatation has no bearing on the severity of pulmonary valve stenosis. The electrocardiogram shows right ventricular hypertrophy (Fig 3); the degree of right ventricular hypertrophy is proportional to the severity of stenosis. Right atrial enlargement may be present. Echocardiogram shows right ventricular enlargement without paradoxical septal motion and thickened and domed pulmonary valve leaflets. The Doppler flow velocity across the site of obstruction is increased and the magnitude of this increase reflects the severity of pulmonary valve stenosis. The peak instantaneous pressure gradient can be calculated by the use of a modified Bernoulli equation:

\[ \Delta P = 4 V^2 \]

where \( \Delta P \) is instantaneous peak pressure gradient in mmHg and \( V \) is the peak velocity across the valve in meters/sec.

It was initially thought that the peak instantaneous gradient is reflective of the peak-to-peak systolic gradient.