Congenital Heart Diseases in Noonan's Syndrome – Mitral Valve Anomalies: Important Cause of Left Ventricular Outflow Tract Obstruction

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Background: Noonan's Syndrome (NS) is a common genetic disorder associated with congenital heart disease. It is an autosomal dominant. Cardiac defects are found in 50-60% of cases of this syndrome. Ventricular outflow obstruction in the form of pulmonary valvular stenosis and hypertrophic cardiomyopathy (HOCM) is the most common congenital heart condition. Apart from HOCM, mitral valve anomalies are also associated with left ventricular outflow obstruction.

Objective: To identify congenital heart diseases associated with Noonan's syndrome.

Design: A Retrospective Study.

Setting: Pediatric Cardiac Outpatient Service, Bahrain Defence Force Hospital, Bahrain.

Method: All patients with confirmed Noonan's syndrome from 2005 to 2018 were included in the study. Cardiac evaluation was performed by chest X-ray, ECG, 2D echocardiography and cardiac catheterization. Personal characteristics were documented: age at diagnosis, sex and consanguinity. Morphologic types of congenital heart diseases (CHD) and outcomes were analyzed.

Result: Twenty-nine children with Noonan's syndrome; 15 (52%) males and 14 (48%) females were included in the study from 2005 to 2018. Pulmonary valvular stenosis, 21 (72%), was the most common cardiac anomaly followed by HOCM, 5 (17%). Mitral valve tissue was found in two (7%) patients. Severe obstruction to the left ventricular outflow by accessory mitral tissue was identified in one (3%) patient which required surgery.

Conclusion: Cardiac defects are common in NS. Left outflow tract obstruction due to accessory mitral valve tissue is rare; however, it is a significant anomaly and if not specifically looked for on echocardiography, it could be missed.

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