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Univentricular CHD: Presentation, management and future expectations

Functionally univentricular hearts represent around 10% of congenital heart defects (CHD). The majority of patients with these conditions do not survive the peopletal period or early in the these conditions do not survive the neonatal period or early infancy without proper intervention. Only a few patients with an optimal balance between pulmonary and systemic circulation may survive into adulthood without surgical intervention. Patients with univentricular CHD represent a group of cardiac malformations almost always determined by a dominant right or left ventricle. The following are the most common subtypes of functionally univentricular hearts: In the first group, the ventricle may present with univentricular atrioventricular connection; the two atrioventricular valves drain into a dominant pumping ventricle as in patients with double inlet ventricle. In the second group, the patients have absence or severe stenosis of either the right or left atrioventricular connection almost always associated with severe hypoplasia of the corresponding ventricle (for example, patients with tricuspid or mitral atresia, hypoplastic left heart syndrome). In the third group, the patients have bilateral atrioventricular connection but either marked hypoplasia of the right or left ventricle or abnormalities of atrioventricular or ventriculoarterial connection precluding a biventricular repair (for example, unbalanced atrioventricular septal defect or complex forms of transposition of the great arteries). So, it is a wide range of congenital heart defects that can be palliated by series of surgical intervention ending with the modified Fontan operation irrespective of the underlying cardiac malformation and associated anomalies of the systemic venous and pulmonary venous return. These groups of patients require lifelong follow up and serial evaluation because, despite reduced operative mortality, there remains a worrying decrement during long term follow up due to sequelae and complications of the univentricular circulation.

Biography

Haitham Talo was the head of Sheikh Khalifah Medical City for 11yrs. He was also working as a pediatric cardiology consultant in Al-Bassel Heart Institute from 2004-2006. Currently he is a pediatric cardiology consultant at Danat Al Emarat hospital, UAE.

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