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Case Report

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Surgical Challenges for Norwood Procedure in Hypoplastic Left Heart with Dextrocardia - A Case Report

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Abstract

Hypoplastic Left Heart Syndrome (HLHS) with dextrocardia is rare and surgical management remains a major challenge. The Norwood procedure is often deferred due to anatomical complexity and hybrid palliative procedure is favored during the neonatal period. We describe our surgical strategy in a baby born with HLHS and dextrocardia. He underwent bilateral Pulmonary Artery (PA) banding and PDA (Patent Ductus Arteriosus) stenting in the neonatal period and subsequent comprehensive Norwood procedure successfully at seven months of age. HLHS with dextrocardia poses specific challenges that can be addressed by stepwise approach. Deferring Norwood by hybrid repair allows better intracardiac repair of associated lesions and better fashioning of DKS (Dmus Kaye Stansel) anastomosis.

Keywords

Hypoplastic left heart syndrome, Dextrocardia, Pulmonary artery banding, Patent Ductus Arteriosus Stenting, Norwood, Damus Kaye Stansel

List of Abbreviations

HLHS: Hypoplastic Left Heart Syndrome; PA: Pulmonary Artery; PDA: Patent Ductus Arteriosus; DKS: Damus Kaye Stansel; AV: Atrio-Ventricular; AVSD: Atrio Ventricular Septal Defect; PAB: Pulmonary Artery Banding; MPA: Main Pulmonary Artery; CPB: Cardiopulmonary Bypass; BT Shunt: Blalock-Taussig Shunt

Introduction

Incidence of HLHS with dextrocardia is rare and its surgical management is particularly complex [1,2]. Comprehensive Norwood procedure in early neonatal period poses specific challenges due to abnormal relationship of great arteries and presence of associated intracardiac lesions in this configuration. A hybrid palliative approach represents a valid option to defer Norwood stage I [3].

Case Presentation

A baby was born at term with antenatal diagnosis of HLHS. Postnatal echocardiography demonstrated dextrocardia, Hypoplastic left Atrio-Ventricular (AV) valve, Hypoplastic left ventricle and hypoplastic arch, unbalanced AVSD (Atrioventricular Septal Defect) without any ventricular component and severe right AV valve regurgitation. There was a left sided Superior Vena Cava and a right sided Inferior Vena Cava to left sided common atrium. A hybrid approach was preferred initially to reduce the surgical burden during neonatal period and to give the baby a trial of life before undergoing a Norwood. Bilateral PA banding (PAB) followed by PDA stenting (Figure 1) was undertaken in neonatal period with a plan for comprehensive Norwood procedure in early infancy. This allowed to defer the Norwood stage I until 7 months of age when the baby had reached 6 Kg.

During Norwood, great vessel arrangement proved to be exceptionally challenging with MPA (Main Pulmonary Artery) coming off anteriorly and crossing horizontally and leftwards and the aortic root and ascending aorta displaced completely

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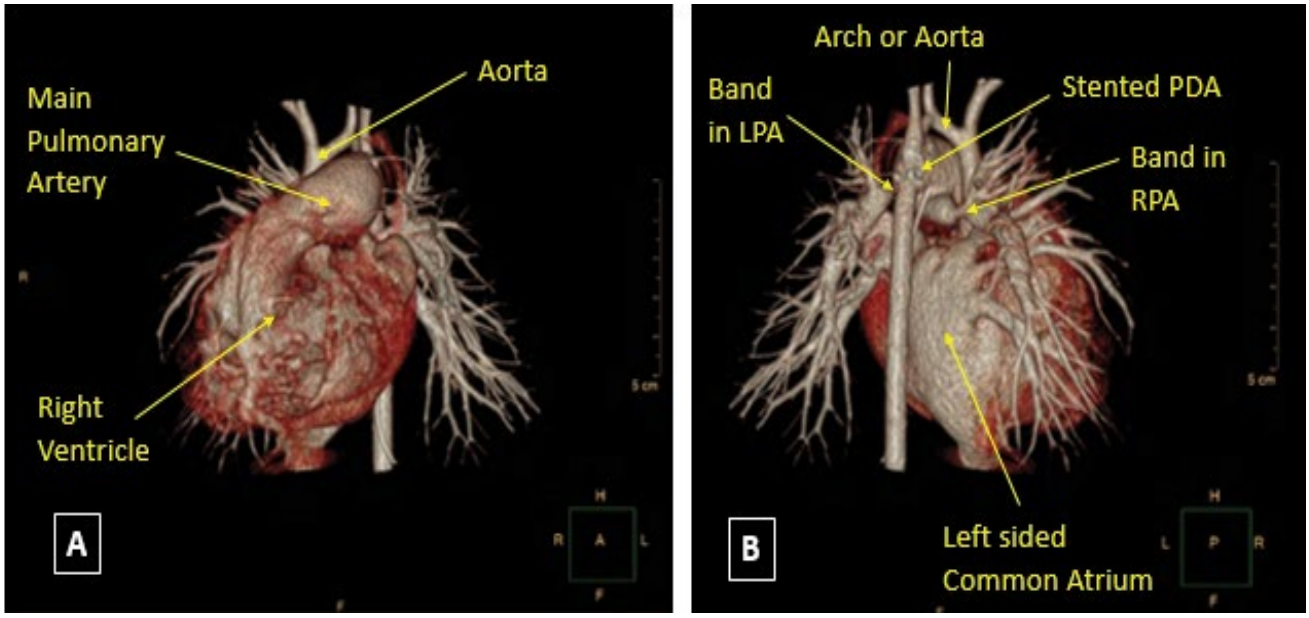

Figure 1: Preoperative images- 3D reconstructed image of CT scan (A) Anterior view showing the relationship of great vessels (Aorta and Pulmonary Artery); (B) Posterior view showing Bilateral PA bands at origin of branch pulmonary arteries, Stented PDA and left sided common atrium receiving pulmonary venous drainage. PA: Pulmonary Artery; PDA: Patent Ductus Arteriosus; LPA: Left Pulmonary Artery; RPA: Right Pulmonary Artery.

posteriorly (Figure 2A and Figure 2B). Branch PAs were taking off in different planes with right PA arising in close relation to pulmonary valve (Figure 1B). Cardiopulmonary bypass (CPB) was initiated with standard ascending aorta and bicaval cannulation. Temperature reduced to 20 degree centigrade. Stented duct was divided in between ligatures. Main PA was divided at pulmonary bifurcation. Both PA bands were removed and branch PAs were dilated with Hegar dilator. Aorta was clamped and cold blood cardioplegia was delivered into aortic root. AV valve was approached through atriotomy and cleft in the right AV valve was repaired.

Arterial cannula was then advanced to right carotid artery, head and neck vessels snared, descending aorta was clamped and selective cerebral perfusion was commenced. Arch reconstruction was undertaken with a homograft patch, however it was felt that DKS anastomosis was going to lie in an unusual orientation. This was addressed by implanting main PA more ventrally than usual to belly of the homograft arch. (Figure 2C and Figure 2D). This allowed completion of anastomosis and reinstitution of full body perfusion and rewarming. Clamp was released at this stage.

Pulmonary circulation was fed via a 4 mm right modified BT (Blalock-Taussig) shunt to right PA which was sitting deeply and posteriorly. This required a second cardioplegic arrest due to difficult access. On weaning from CPB, there was profound desaturation, hence decision was made to add a 6 mm Sano shunt to left sided pulmonary bifurcation. This allowed weaning off the cardiopulmonary bypass keeping both shunts open. BT shunt was eventually clipped. Successive echocardiography demonstrated good ventricular function, good flow in branch pulmonary arteries and mild regurgitation across atrioventricular valve. CT scan performed subsequently showed good surgical result (Figure 3).

Discussion

HLHS with dextrocardia and associated intracardiac defects are rare [1]. Anatomical complexity and relationship of great vessels poses specific challenges and surgical repair should be individualized.

We used a stepwise approach to address specific issues. Although the outcome after Norwood procedure in average risk patients have significantly improved [2], in patients with specific risk factors like prematurity, intact atrial septum, severe valve insufficiency, low birth weight (< 2.5 Kg), genetic abnormality, interstage hospital mortality is 40-60% [3]. Early result after hybrid palliation with ductal stenting and bilateral PA banding for such patients are encouraging [4] and long-term survival are comparable to those who undergo primary Norwood procedure [5]. Arrangement of the great vessels in dextrocardia poses problem in fashioning DKS anastomosis without distorting neo-aortic valve. This was addressed in our case by implanting the PA more posteriorly onto the under surface of the reconstructed arch. Moreover, due to rotation of heart, right PA was deeper than in usual arrangement making insertion of right sided BT shunt technically challenging. Sano shunt is probably a better option to consider in such scenarios due to technical ease and better disposition to achieve good pulmonary flow. In literature, 12 months transplant free survival have been reported to be better after Sano shunt than Modified BT shunt [6].

To summarise, HLHS with dextrocardia poses specific challenges that can be addressed by stepwise approach. Deferring Norwood by hybrid repair allows better intracardiac repair of associated lesions and better fashioning of DKS anastomosis.

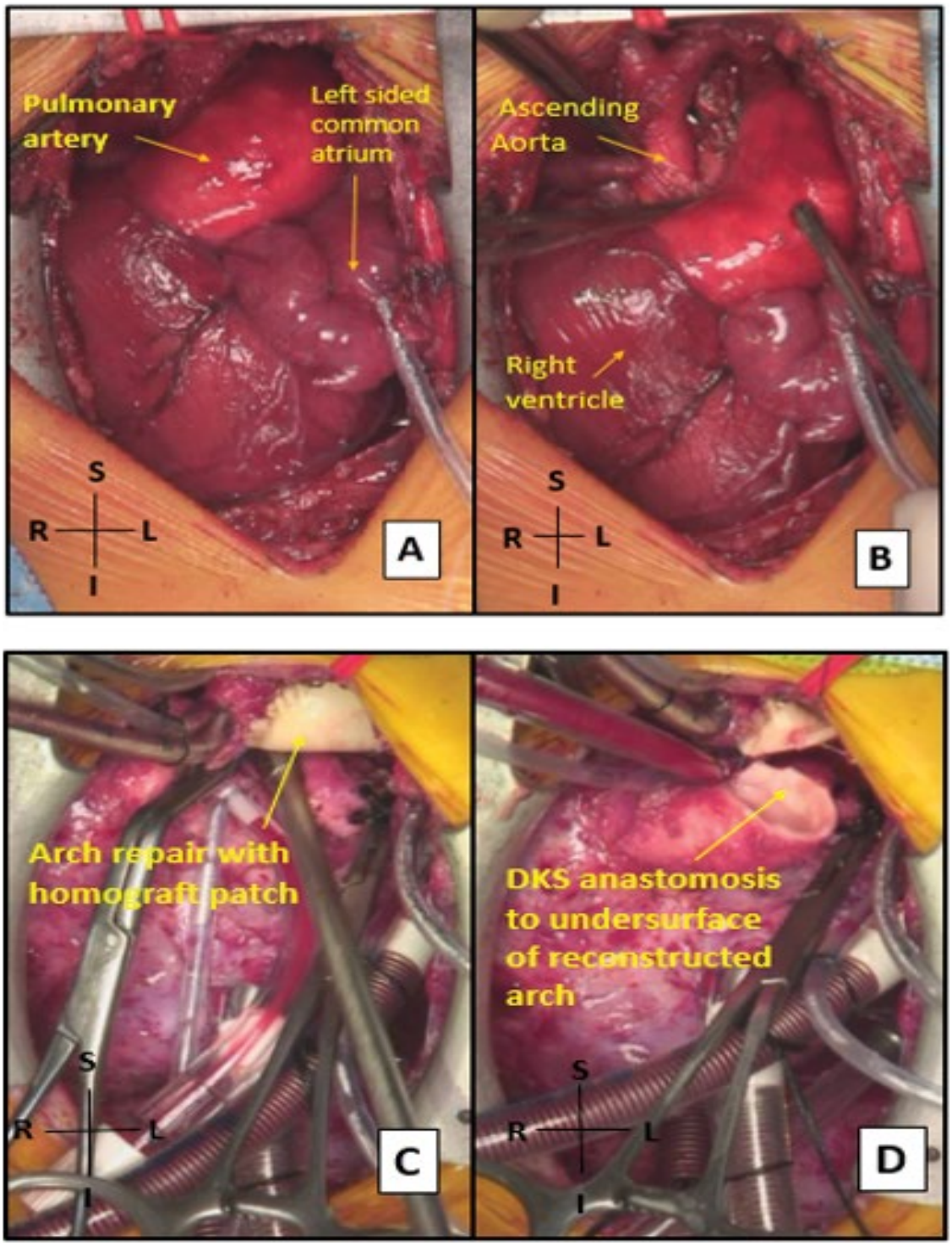

Figure 2: Intraoperative images. Preoperative anatomy showing (A) Dextrocardia, left sided common atrium with Main Pulmonary Artery crossing left to right in front of Aorta; (B) Main Pulmonary artery retracted towards left to show hypoplastic ascending aorta and arch of aorta; (C) Repair of hypoplastic ascending aorta and arch of aorta with homograft patch; (D) DKS anastomosis- anastomosis of Main Pulmonary Artery to undersurface of belly of homograft arch. DKS: Damus Kaye Stansel.

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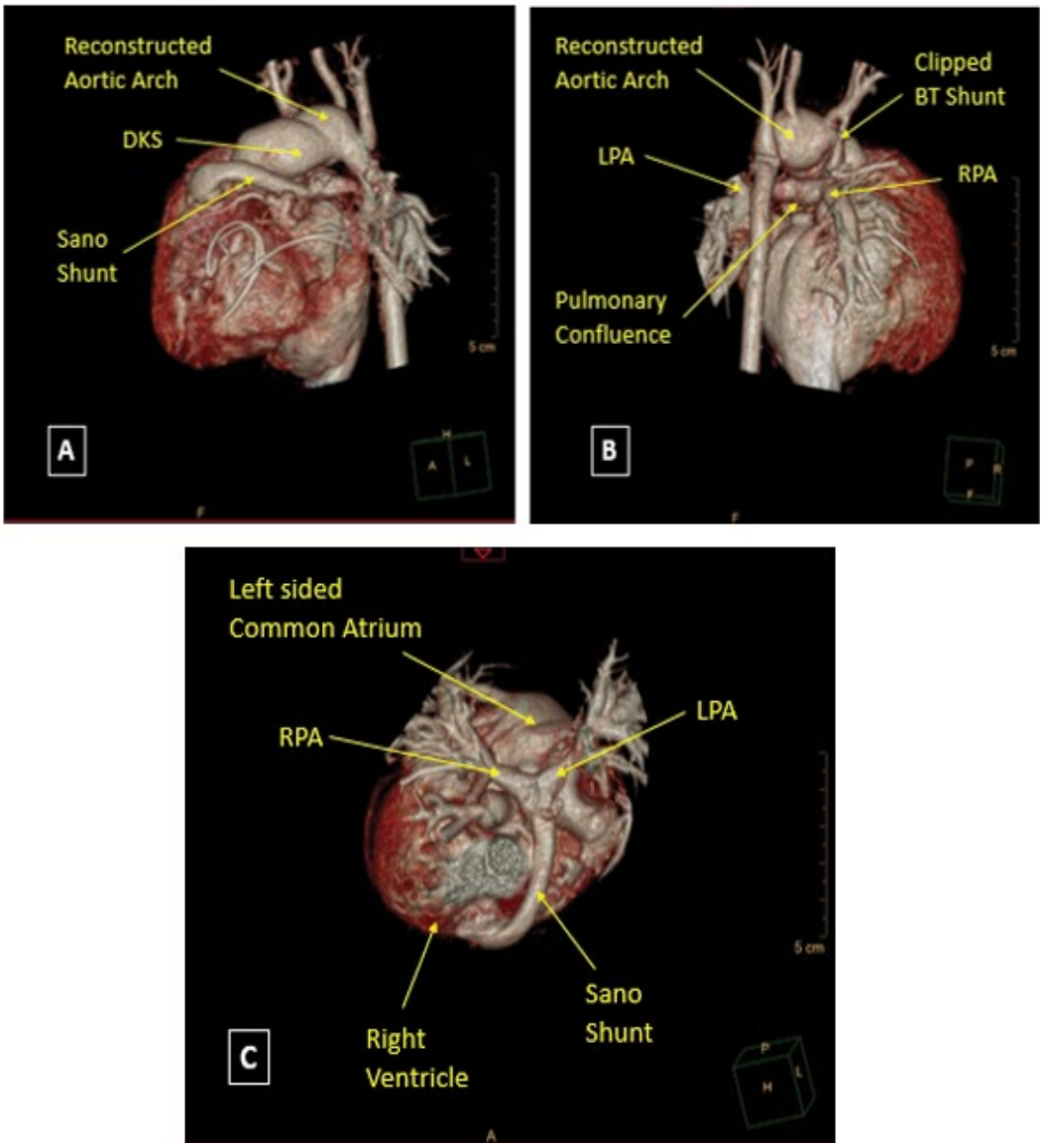

Figure 3: Postoperative images. 3D reconstructed image of CT scan (A) Left Anterolateral view showing DKS, reconstructed arch of aorta and Sano shunt; (B) Posterior view showing pulmonary confluence with branch pulmonary arteries and reconstructed aortic arch; (C) Cranio-caudal view showing Sano shunt with adequate pulmonary confluence and good-sized branch pulmonary arteries. DKS: Damus Kaye Stansel.

Conflicts of Interest

The author(s) declare no conflicts of interest.

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