Considerations in biventricular repair after the Norwood procedure

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Abstract

Objective: The Norwood procedure can be applicable as a first stage palliation in children who can eventually undergo a biventricular repair. Although usual management of these patients is a primary neonatal repair, in selected patients staged approach with a Norwood procedure in the neonatal period followed by a Rastelli procedure in the infancy for conversion to two-ventricle physiology has been used alternatively. Methods: We report our experiences on two infants who underwent a previous palliation with the Norwood procedure for lesions other than hypoplastic left heart syndrome and converted to two-ventricle physiology by the use of a Rastelli-type procedure. This report examines considerations in biventricular repair after the Norwood procedure especially need for ventricular septal defect enlargement and approach to placement of the right ventricle to pulmonary artery conduit. Results: Both of the infants who underwent staged approach with an initial Norwood procedure for lesions other than hypoplastic left heart syndrome survived the operations and were clinically well at mid-term follow-up. Conclusion: In selected patients, the staged approach is an alternative in management of malformations other than hypoplastic left heart syndrome which share the important physiologic features of aortic outlet obstruction and ductal dependency of systemic circulation. We recommend routine enlargement of ventricular septal defect and proper positioning of the conduit at the time of subsequent biventricular repair. © 2000 Elsevier Science B.V. All rights reserved.

Keywords: Critical left ventricular outflow tract obstruction; Staged approach with an initial the Norwood procedure; Biventricular repair

1. Introduction

The Norwood procedure was originally designed and used as a first stage palliation in patients with hypoplastic left heart syndrome in the pathway leading to an eventual Fontan procedure. However, the Norwood procedure can also be used as a first stage palliation in newborns with a variety of other malformations other than hypoplastic left heart syndrome which share the important physiologic features of aortic outlet obstruction and ductal dependency of systemic circulation. Heart defects which may be treated by the latter approach include; (1) interrupted aortic arch and ventricular septal defect (VSD) with severe subaortic stenosis, (2) aortic atresia and VSD with normal sized left ventricle, (3) double outlet right ventricle with subaortic stenosis and aortic arch hypoplasia. Two approaches can be applicable in surgical management of that kind of lesions; biventricular repair in the neonatal period or initial palliation with Norwood procedure and subsequent biventricular repair with the use of Rastelli-type procedure. In the
2. Case reports

2.1. Case 1

The patient was born, at 37 weeks gestation weighing 2.8 kg. Preoperative studies including echocardiogram and cardiac catheterization revealed interrupted aortic arch (IAA) type B, aberrant right subclavian artery, large malalignment VSD, bicuspid aortic valve, severe subaortic stenosis, patent ductus arteriosus and good biventricular function. Measurements were as follows, subaortic area 3 mm, ascending aorta 4 mm, transverse aortic arch 2 mm and descending aorta 6 mm. Because the results of the primary repair has been reported unfavorably in infants who have subaortic area 3 mm or less [1,2], a staged approach was planned. On the ninth day of life, patient underwent a Norwood procedure with repair of interrupted aortic arch. The cephalic aspect of the aortic arch was directly re-anastomosed and the undersurface of the arch was augmented with pulmonary homograft incorporating the native pulmonary valve proximally as the neoaoartic valve. The atrial septum was excised. A 3.5 mm Gore-Tex graft shunt was placed between the innominate and right pulmonary artery. The postoperative course complicated with severe cyanosis due to central posterior–anterior (PA) stenosis, an additional 3.5 mm Gore-Tex graft shunt between aorta and left PA was placed.

Cardiac catheterization, at age 7 months, showed two good-sized ventricles with good function, no obstruction in the reconstructed ascending aorta and aortic arch, the single posterior malalignment VSD and moderate subaortic stenosis measuring 5.5 mm. There was a moderate to severe obstruction of the confluence of the pulmonary arteries with patency of the two shunts supplying the right and left pulmonary arteries, respectively. By the age 2 years, systemic oxygen saturation had decreased and biventricular repair was undertaken utilizing neoaoartic and bicaval cannulation, a posterior malalignment VSD was exposed via right ventriculotomy. Subaortic area appeared too small to allow adequate resection to relieve the obstruction. VSD appeared mildly restrictive and enlarged by resecting its left and anterior border. VSD was then routed with an intraventricular baffle to pulmonary (neoaoartic) valve. The central shunts were taken down and the central pulmonary arteries opened across the stenosis and anastomosed to the distal end of a 20 mm pulmonary homograft. The distance from the right ventriculotomy to pulmonary arteries was long and necessitated proximal extension of the homograft with a ringed Gore-Tex tube graft. To avoid compression by the sternum, the right ventricle to pulmonary artery conduit was routed to the left side of the neoaoarta. Direct needle measurement of the pressures prior to chest closure showed no gradient between the left ventricle and ascending aorta. Postoperative echocardiography showed normal ventricular function, mild mitral and tricuspid insufficiency, no residual ventricular septal defect and no left ventricular outflow tract obstruction. The patient was clinically well 2 years after operation with the similar echocardiographic findings.

2.2. Case 2

The patient was born at 37 weeks gestation, weighing 2.2 kg. Echocardiogram revealed mesocardia, double outlet right ventricle, subpulmonic VSD, side-by-side great arteries, anterior deviation of the infundibular septum with subaortic stenosis, aortic arch hypoplasia and/or coarctation of the aorta. Measurements were as follows, subaortic area 4 mm, ascending aorta 5 mm and aortic arch 3 mm. Considering the side-by-side relationship of great arteries and the potential problem to place a pulmonary artery-right ventricle conduit in a newborn with mesocardia, primary repair was not planned. On the seventh day of life, the patient underwent a Norwood procedure, with pulmonary allograft augmentation of the aortic arch, proximal association of the aorta and pulmonary valve, atrial septectomy and placement of a 3.5 mm Gore-Tex right modified Blalock–Taussig shunt.

Follow-up cardiac catheterization at age 6 months showed, mesocardia, double outlet right ventricle, single subpulmonic VSD, side-by-side great arteries, anterior deviation of the infundibular septum with dynamic subaortic stenosis, two good sized ventricles with good function, widely patent neo-aorta without any gradient, patent modified Blalock–Taussig shunt. Arterial oxygen saturation ranged from 75 to 85% over next ten months. At age 16 months, the patient underwent biventricular repair. The neoaoarta was large and the branch pulmonary arteries lay far posteriorly and somewhat was compressed behind to aorta. Utilizing neoaoartic and bicaval cannulation the VSD was exposed via a right ventriculotomy and enlarged by resecting its left and anterior borders. VSD was then routed with an intraventricular baffle to pulmonary (neoaoartic) valve. The subaortic area which was diminitive closed with a separate patch. Central shunt was taken down. Once again, it appeared that sternal compression of the right ventricle to pulmonary artery connection could be best avoided by routing a conduit to the left of the neoaoarta. An end-to-side anastomosis was sewn between the end of a 16-mm aortic homograft and central pulmonary arteries. (The anastomosis was sewn to the right of the neoaoarta for convenience. The homograft was then passed underneath the neoaoarta and brought leftwards and up to the ventriculotomy). The back wall of the proximal homograft was sewn to the leftward lip of the ventriculotomy and a Gore-Tex patch to roof the anastomosis. After coming off bypass direct needle measurement showed no gradients between left ventricle and aorta or right ventricle and conduit. Due to remaining concern over conduit compression by the sternum, the posterior table of the sternum was resected prior to chest closure. The patient was clinically well 18 months after operation and a echocardiography study showed normal biventricular function, no residual
ventricular septal defect, no left ventricular and right ventricular outflow tract obstruction. The essential steps in the surgical procedure are illustrated in Fig. 1.

3. Discussion

There exist two ways of surgical management of neonates with good sized two ventricle, severe subaortic stenosis accompanying hypoplastic or interrupted aortic segments with increased pulmonary blood flow; biventricular repair in the neonatal period or palliation with Norwood operation and subsequent biventricular repair with the use of Rastelli-type procedure. The posterior displacement of infundibular septum is the main cause of subaortic stenosis in neonates with VSD and aortic arch obstruction [8]. This situation is almost the opposite of that seen in tetralogy of Fallot in which the anteroaerial displacement of infundibular septum results in right ventricular outflow tract obstruction. The most difficult issue in repair of aortic arch interruption with VSD is the issue of the subaortic obstruction. It is not readily possible to identify those patient preoperatively who have significant subaortic obstruction by either echocardiography or catheterization, since most flow goes across the VSD to the pulmonary arteries preoperatively. This is also the case for patients who have undergone a previous Norwood operation, at the time of biventricular repair. Bowe and associates [2] has reported excellent results with neonatal anatomic repair combining reconstruction of the aortic arch, resection of infundibular septum and closure of VSD on small group of newborns with IAA-VSD and severe subaortic obstruction. In these patients, if the subaortic stenosis can not be relieved by resection of the infundibular septum, a complex operation combining the features of the Norwood procedure and the Rastelli procedure which constitutes establishing a continuity between proximal main pulmonary artery and aorta, baffling the VSD to the pulmonary artery with a patch and achieving right ventricular-pulmonary artery continuity with a homograft conduit can be applicable [9]. However, in selected patients, the staged approach has been an alternative for surgeons. The first approach has the advantages of establishing normal physiology from the beginning of life and the avoidance of period of left ventricular volume load due to systemic to pulmonary shunt. Two-stage approach however, avoids the very complex operation and potential problems with pulmonary artery-right ventricle conduit replacement in the neonatal period especially in a small newborn with unusual cardiac configuration such as mesocardia. Even though the results of the Norwood procedure have improved especially for patients with lesions other than hypoplastic left heart syndrome [5–7], the staged approach has theoretical disadvantage of two high-risk procedures in neonatal and infancy period instead of one. Additionally, staged approach exposes the patient with potential long-term problems such as right ventricle to pulmonary artery conduit occlusion and pulmonary valve in systemic position (Fig. 1).

The subaortic obstruction in the hearts with Taussig–Bing malformation is a known entity. In the most of the cases the obstructive lesion is mainly a restrictive VSD [10,11], however, it may be hypertrophied infundibular septum [12] or double straddling of the tricuspid and mitral valves [11].

In children with double outlet right ventricle with severe subaortic stenosis with subpulmonic VSD and aortic arch obstruction, surgical management depends on the relationship of great arteries. Primary biventricular repair can usually be applicable in patients with anterior–posterior relationship of great arteries with resection of infundibular septum, construction of a VSD closure connecting the left ventricle to pulmonary artery followed by an arterial switch procedure and repair of aortic arch obstruction. In the configuration of side-by-side relationship of great arteries, if the minimal tricuspid-pulmonary valve distance is greater than the aortic size, although an arterial switch procedure is always feasible, Serraf and associates have advocated an intraventricular repair after a period of palliation with pulmonary artery banding [13].

In infants who has undergone a previous Norwood operation with good sized two ventricle, usual management is conversion to two-ventricle physiology by the use of Rastelli procedure unless there are accompanying uncorrectable malformations such as straddling atroventricular valves [5–7,14]. This operation is usually performed between the age of 18–24 months. The other option for these patients is continuation in univentricular palliation with a fontan completion. A recent report from Great Ormond Street, investigating the applicability of single-ventricle repair in children with complex heart defects and good sized two ventricles, has shown comparable results to biventricular repair in an average 3-year follow-up [15]. However 3-year follow-up period may not be long enough to reach a conclusion.

Successful conversion to two-ventricle physiology by the use of Rastelli-like procedures has been reported in these patients [5–7,14]. In technical point of the view, some of the important points include the enlargement of VSD to prevent the development of potential subneoaortic stenosis and positioning the right ventricle-pulmonary artery conduit in order to avoid sternal compression.

Prediction of the postoperative development of left ventricular outflow tract obstruction due to a restrictive VSD may be difficult. After Norwood procedure because of atrial septectomy, only some of pulmonary venous return may pass via left ventricle. Therefore, it is usually hard to predict gradient across the VSD. After Rastelli procedure, because atrial septum will be no longer open, entire cardiac output must pass through VSD. This change may reveal some degree of restriction in blood flow through VSD in the early postoperative period. A modification in the Norwood procedure without doing an atrial septectomy
may be suggested for the patients with the lesions of good sized two ventricles. Leaving a restrictive atrial septal defect (ASD) may result in well-growth left ventricle and more accurate assessment of VSD gradient before conversion to biventricular repair.

Development of subaortic stenosis after biventricular repair of double outlet right ventricle particularly in Taussig–Bing malformation has been previously reported. The obstruction usually originates from a restrictive VSD [10,11,16]. VSD enlargement has been advocated, before patch closure, to prevent subaortic obstruction development [11,12,17]. Similarly, obstruction at the subneoaoartc area may occur after tunneling left ventricle through a restrictive VSD to the neoaoaorta. Routine enlargement of the VSD may be suggested during Rastelli operation in children who had a previous Norwood procedure. VSD enlargement is usually done with resection of its anterocephalic rim through conal and parietal structures to preserve conduction system as previously described [11,17].

After the Norwood operation, due to the substantial size of the neoaoaorta, positioning of the right ventricle to pulmonary artery conduit is an important point. The location of pulmonary arteries posteriorly behind the neoaoaorta limits the space and increased the distance for the conduit. It is
usually easier to route the conduit to the left rather than to the right. Especially in hearts with mesocardia, because the right ventricle lies directly behind the sternum, doing a low right ventriculotomy and using a pulmonary homograft conduit extended proximally by ringed Gore–Tex tube graft may be advisable to give the conduit a large curve over the left side of the heart. Beveling polytetrafluorethylene (PTFE) extension as a hood should make the proximal anastomosis to the ventricle [18]. Even the conduit can be kept away from sternum, it may be compressed underneath the neoaorta at the anastomosis site to the pulmonary arteries. This point should be remembered if unexplainable high pulmonary homograft conduit pressure measured after coming off by-pass. Some other preventive measures such as to open the left pleura all the way down to let the heart deviate left hemithorax and resection of the posterior table of the sternum should be taken. Direct needle measurement of the right ventricle pressures after sternal closure is also advisable to make sure that there is no elevation due to compressed conduit.

In conclusion; in selected patients, the staged approach is an alternative in management of malformations other than hypoplastic left heart syndrome which share the important physiologic features of aortic outlet obstruction and ductal dependency of systemic circulation. However, biventricular repair after the Norwood procedure may be complicated by obstruction at the subneoaortic area due to a restrictive VSD and compression of the right ventricle to pulmonary artery conduit due to anterior large neoaorta. Routine enlargement of the VSD and proper positioning of the conduit may prevent these problems.

References


