Emergency Starnes Operation: A Surgical Success Beyond Any Better Expectation

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Ebstein’s anomaly is a rare malformation of the tricuspid valve representing 0.5% to 1.0% of all congenital heart disease and consisting of failure of the delamination of the valve leaflets from the endocardium, resulting in variable degrees of dysplasia and downward displacement of the posterior and septal leaflets of the tricuspid valve [1–8]. The anomalous valve is often regurgitant; the inlet portion of the right ventricle (RV) becomes atrialized, and an interatrial communication is usually present, facilitating a right-to-left shunt (Fig 1) [1, 2]. Ebstein’s anomaly may show various forms of presentation, from severely symptomatic neonatal forms (with severe tricuspid regurgitation and functional pulmonary atresia) to an asymptomatic form incidentally discovered late in adulthood [8].

In 1991 Starnes and colleagues [2] introduced a new palliative procedure in 5 critically ill neonates with Ebstein’s anomaly presenting with cyanosis, acidosis, and congestive heart failure. The procedure consisted of tricuspid closure with autologous pericardium and an aortopulmonary shunt of 4-mm polytetrafluoroethylene tubing. Exclusion of the RV has been subsequently extended to older patients with severe Ebstein’s anomaly [5] and to other diseases such as arrhythmogenic RV dysplasia causing intractable heart failure [7].

A 6-year-old boy came to our attention during our medical humanitarian mission in the cardiothoracic unit of the International Operating Centre for Children in Asmara (IOOCA) in Eritrea, run by ArchiMED, a German nongovernmental charity organization (www.archimed.org). The boy, embraced by his father, was in agony after a 300 km troublesome and exhausting journey across the country from a rural village to the capital. The child was severely cyanotic (arterial oxygen saturation 60%) and dyspnctic, with weak central pulses and poor peripheral perfusion. He had an immensely enlarged liver, ascites, head, neck, and peripheral edema, tachycardia, and a systolic murmur. He was dehydrated and malnourished, and had hypoproteinemia and polycythemia (with an hematocrit of 68%).

An echocardiogram performed in the emergency room revealed Ebstein’s anomaly with a severely dysplastic, deeply downward displaced tricuspid valve causing severe regurgitation, and huge enlargement of the right atrium and of the atrialized portion of the RV. The Great Ormond Street echocardiography (GOSE) score (ie, area of the right atrium plus area of the atrialized portion of RV divided by area of the RV plus area of the left atrium plus area of the left ventricle [LV]) was 3 to 4 [1]. There was a patent foramen ovale with an exclusive right to left shunt, a dysplastic pulmonary valve with mild stenosis, and mild insufficiency. The LV was compressed by important interventricular septal bulging that was impinging on its hemodynamic function.

The boy was promptly intubated, sedated, put under mechanical ventilation, and prepared for surgery. Considering the unfavorable anatomy of the tricuspid valve and the severe general condition of the child, a decision was taken to operate immediately using a lifesaving procedure. The operation consisted of closing of the tricuspid valve with a patch, enlarging of the atrial septal defect, and establishing a cavopulmonary connection (bidirectional Glenn) under cardiopulmonary bypass. Cardiopulmonary bypass time was 56 minutes, and the procedure was accomplished at 30°C nasopharyngeal temperature. According to our previous experience with neonatal Starnes operations, we decided not to fenestrate the tricuspid patch and to leave the pulmonary
valve untouched. The operation was complicated by complete atrioventricular block. An external pacemaker was used as a bridge until permanent epicardial wires and an implantable pacemaker were sent from Europe and implanted 7 days after the operation.

The postoperative course was uneventful; the patient was extubated the morning after operation (maintaining a good oxygen saturation of 85%), and a surprisingly progressive and rapid improvement of both hemodynamic and clinical status occurred over the following days. The echocardiographic findings showed a well-functioning cavopulmonary anastomosis, mild pulmonary insufficiency, no more signs of interventricular septal bulging to the LV, and good LV systolic function (ejection fraction calculated by four-chamber view was 65%). The RV, due to unloading, had a marked reduction of its cavity. The boy was discharged on the 15th postoperative day owing to the need for pacemaker implantation.

At 1 year follow-up, the boy was in good general condition, well compensated, and fully active, with a mixed venous oxygen saturation of 78%. At echocardiography, moreover, the RV cavity had become virtually minute (Fig 2). On the roentgenogram, the heart was not enlarged, and the child also recovered his sinus rhythm. The child is at the moment waiting for completion of Fontan circulation.

Comment
In 1991, Starnes and colleagues [2] described a new surgical palliative procedure for neonates with Ebstein’s anomaly consisting of tricuspid valve closure with autologous pericardium and an aortopulmonary shunt with a 4-mm polytetrafluoroethylene graft. The Starnes operation is usually reserved for neonatal age patients [2-4] and is accomplished by closure of the tricuspid valve and a systemic to pulmonary shunt. Nonsignificant differences have been reported in fenestrated versus nonfenestrated patches, although fenestration is generally preferred [4, 6]. Total RV exclusion with total cavopulmonary connection may be an option in adult patients [5-7]. Reports on modified Starnes procedures in childhood and including partial cavopulmonary connection are, however, very limited [6].

The decision to perform a bidirectional Glenn rather than an aortopulmonary shunt was due to the patient’s age and to the high incidence of shunt occlusion encountered in our experience in this high sea level country. We decided not to fenestrate the patch because the risk of thromboembolic events was considered significant owing to the high altitude, the inherent risk of dehydration in this sub-Saharan country, and the impossibility of managing anticoagulant drugs outside its capital Asmara. A certain concern for the future enlargement of the RV, and its deleterious effects on LV function, remained because the pulmonary insufficiency and stenosis, albeit mild, could contribute to RV dilation. At follow-up 1 year after the operation, the concerns have cleared away because the RV cavity had become virtually minute, with no signs of compression of the LV.

Probably in this case, other than unloading, the RV could have been further decompressed by a patent pulmonary valve. The peculiarities of this case are the dramatic clinical and geographic scenario, and the rapid and surprising postsurgical improvement. The RV exclusion in combination with a bidirectional Glenn procedure allowed for unloading of the right side of the heart with

Fig 1. Thorax radiograph (A) before, (B) immediately after, and (C) 1 year after intervention showing an enormous cardiomegaly that progressively resolved after surgery.

Fig 2. Postoperative echocardiographic image in four-chamber view showing the right ventricle (RV) cavity had become virtually minute, with no signs of compression of the left ventricle (LV). (LA = left atrium; RA = right atrium.) (The image’s quality is consistent with the technology available in a Third World country.)
rapid resolution of right-side heart failure and subsequent amelioration of LV function.

Extreme cases of Ebstein’s anomaly like the one described in this case report are rare in developed countries, whereas they may more often present to a surgical team when working in a humanitarian mission in Third World countries. The choice to perform very high risk surgery within a setting of limited resources (and material) against a decision to leave such cases untreated is challenging, not to mention the ethical set of problems behind it. To operate on this case was clearly a gamble, but to wager and battle for the sake of a sick patient with no other options is a worthwhile and sometimes rewarding task.

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References

Leiomyosarcoma of the Superior Vena Cava

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Leiomyosarcoma of the superior vena cava is a very rare tumor and only a few cases have been reported, with various techniques of vascular reconstruction. We describe a new case of leiomyosarcoma of the superior vena cava in a 61-year-old woman with extension to the brachiocephalic arterial trunk. Resection and vascular reconstruction were performed using, respectively, polytetrafluoroethylene and polyethylene terephthalate vascular grafts.

Vascular leiomyosarcoma is a very rare tumor and the inferior vena cava is the most common site (50%) [1]. The superior vena cava (SVC) is exceptionally involved and only a dozen cases have been previously reported [2]. The tumor is commonly revealed by physical signs of a SVC obstruction syndrome, less frequently by metastasis or unexpected computed tomography scan finding. Leiomyosarcoma is considered a very locally aggressive tumor with rare distant dissemination and an...