Commentary: Ebstein’s Anomaly with circular shunt – round and round we go but not necessarily very fast.

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Commentary: Ebstein’s Anomaly with circular shunt – round and round we go but not necessarily very fast.

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Central Message: Neonates with Ebstein’s Anomaly and circular shunt need to be differentiated into those who are hemodynamically unstable (a surgical emergency) and those who are hemodynamically relatively stable.

Central Picture Legend: Christopher J Knott-Craig MD FACS

Konstantinov and colleagues are to be congratulated on the successful neonatal biventricular repair of a patient with Ebstein’s Anomaly (EA) and a circular shunt (1). These are very challenging patients with historically high hospital mortality rates of around 40-90% (2,3).

The first successful repair of a neonate with EA and hemodynamically unstable circular shunt was done in 1994 and published with 5-year follow-up by our group in 2000 (4). This patient is currently asymptomatic, married with a family and fully employed 29 years later. Due to the perceived high mortality associated with neonates with EA and circular shunt, there is currently increased advocacy for initially doing a single ventricle palliation (Sternes’ operation) (5) and deferring bi-ventricular conversion to late infancy, as successfully done several times by Drs Jose and Luciana Da Silva in Pittsburg; (6). It should be noted that this bi-ventricular conversion was also first done by our group in Oklahoma in 2000 on a 2-month-old ventilator –dependent patient who had undergone a Starnes’ operation elsewhere (7). In that case, as in the case reported by Konstantinov in the current issue of JTCVS, the patient had associated RVOTO; this allowed the surgical team to obtain a tricuspid valve regurgitant pressure gradient which in both cases was >40 mm Hg, predicting a suitable right ventricle for successful biventricular repair.
Neonates with EA and circular shunt constitutes a spectrum of clinical scenarios: on the one hand, in our experience, most symptomatic with neonates and functional pulmonary atresia initially have some pulmonary regurgitation arising from the patent ductus flow directed at the pulmonary valve in the context of neonatal elevated pulmonary vascular resistance. And since these neonates have moderate to severe tricuspid valve regurgitation, this technically constitutes a “circular shunt”. These neonates are candidates for a Starnes’ operation, but are also often suitable for an initial bi-ventricular repair with an expected mortality of around 10% (9).

On the other hand, neonates with EA and circular shunt who are critically unstable including those with associated cardiogenic shock constitutes an altogether different subgroup of patients; these need immediate surgical intervention aimed at interrupting the circular shunt as advocated by Konstantinov and colleagues. The surgical options include immediate Starnes’ operation, or temporary ligation of main pulmonary artery and placement of bilateral pulmonary artery bands on the branch PAs followed by early Starnes’ operation or bi-ventricular repair once the end-organs have recovered, depending on the morphology present.

Once again, the authors are to be congratulated on an excellent outcome with 2-year follow-up.

References


