Management of neonatal Ebstein’s anomaly: towards a rational approach?

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Sano and collaborators report a small series of 12 neonates and infants who underwent surgery for Ebstein’s anomaly. The results are outstanding, with 10 survivors (83%) after a mean follow-up of 6.5 years [1].

It is well recognized that Ebstein’s anomaly of the tricuspid valve is much more than just a valvar malformation because the condition involves the structure and function of the entirety of the right heart. Symptomatic neonates present with a combination of cyanosis (due to right-to-left shunting and/or decreased pulmonary blood flow) and congestive heart failure (due to tricuspid regurgitation and/or right ventricular dysfunction). The management of neonatal Ebstein’s anomaly has been controversial. There is a general agreement that aggressive medical treatment should be offered first and that a significant proportion of the population (approximately half of the patients) stabilizes and improves progressively as the pulmonary vascular resistances decrease. There remains, however, a considerable debate regarding those neonates who fail weaning from medical support and need surgical treatment. An initial one-ventricle approach is often advocated [2], whereas bi-ventricular repair is favoured by other groups [3].

The present paper offers useful information regarding several points of discussion.

(i) It provides confirmation that a majority of neonates with Ebstein’s anomaly can safely undergo a complete two-ventricle repair (7/12 patients, without mortality, in the present series). The essential principles of bi-ventricular repair include: (1) creation of a competent tricuspid valve, (2) plication of the atrialized part of the right ventricle, (3) reduction of the right atriotomy and (4) relief of right ventricular outflow tract obstruction if present. Various techniques have been used to repair the tricuspid valve. Cone reconstruction is gaining wide acceptance for older children and adults; because of its effectiveness and reproducibility, it may become the procedure of choice in neonates and small infants as well [3, 4].

(ii) There are, however, patients in whom the right ventricle is unable to sustain, even in part, the cardiac output; there obviously remains a role for single-ventricle palliation. The first surgical step includes right ventricular exclusion by patch closure of the tricuspid valve. To avoid distension of the right ventricular cavity, with subsequent abnormal bulging of the inter-ventricular septum into the left ventricular outflow tract, it is mandatory to leave a fenestration in the patch. As suggested by Sano and collaborators, another more radical approach to this issue is to associate closure of the tricuspid valve with resection of the free wall of the right ventricle. Although the experience in 3 patients is encouraging (three early survivors, one late death after Fontan completion), further experience is necessary before recommending this approach. Another potential solution is to excise the tricuspid valve, close the right ventricular outflow tract and leave the right ventricular cavity as a non-functioning atrialized chamber.

(iii) The evaluation of the right ventricular function represents the key point of the decision making. Several factors should be taken into consideration: size and morphology of the non-atrialized portion of the right ventricle, antegrade pulmonary blood flow, and, as suggested by Sano and collaborators, ability of the right ventricle to generate pressure (as evaluated by the trans-tricuspid systolic pressure gradient). Because of the rarity of the malformation, it is impossible for individual centres to accumulate large series. I suspect that, in many cases, the decision is made subjectively by guesswork. Multicentric studies are necessary to define better objective criteria. Meanwhile, it seems reasonable to favour bi-ventricular repair and to attempt valve repair in the majority of neonates with Ebstein’s anomaly, before converting them to single-ventricle physiology.

REFERENCES


