The challenging quest of neonatal Ebstein management

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Ebstein anomaly of the tricuspid valve represents a spectrum of abnormalities affecting the development of the right heart, which can be associated with variable pathophysiology, presentation (Figure 1), and management, depending on age. Management of symptomatic neonates with Ebstein anomaly constitutes one of the most significant challenges surgeons who specialize in congenital heart surgery are confronted with these days. If we consider the rarity of this lesion, the challenging physiology, and the important gaps in knowledge, it is not surprising that management choices are multiple, with variable success, leaving considerable opportunity for improvement.

In this light, the need for appropriate data to inform the complex decisions necessary to optimize the management of these newborns is essential. This need carries even more significance in an era in which decisions about termination of pregnancy in patients with this lesion are based on prenatal assessment without a robust understanding of the natural history and the interplay of the anatomic and physiologic variables.

When one encounters a neonate with Ebstein anomaly, 2 essential questions need to be answered: (1) Can this be managed medically? (2) If a surgical intervention is required, is a biventricular repair feasible now or in the future? The analysis published by Baek et al¹ in this issue of the *Journal* is focused on the second question.

A number of variables play an important role in this decision, namely the degree of tricuspid regurgitation and the possibility of ameliorating this by virtue of allowing the pulmonary vascular resistance to decrease over time or by means of reconstructing the tricuspid valve, the function of the right ventricle and its ability to generate pressure, and not least the patency of a right ventricular outflow and the pulmonary valve. Ideally, antegrade pulmonary blood flow could be established with or without an intervention to ameliorate the degree of tricuspid regurgitation. Alternatively, the negative impact of an atretic pulmonary valve when attempting a biventricular repair in a newborn has been demonstrated clearly. Therefore, the need is imperative to make a distinction between a pulmonary valve that would be able to allow antegrade flow

Neonatal Ebstein.

Central Message

The importance of right ventricular outflow/pulmonary valve complex in the management of neonatal Ebstein is discussed.

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once pulmonary vascular resistance decreases and one that will not.

To this effect, Baek et al¹ evaluated features of the pulmonary valve anatomy to identify those neonates who despite presenting without antegrade pulmonary blood flow could potentially establish it over time, allowing a biventricular or at least one and half ventricular circulation. Although the numbers are small, several conclusions can be drawn from this experience. In the presence of a normal pulmonary valve, a biventricular circulation can be accomplished in the majority of the cases. Pulmonary regurgitation, which was present in all of these patients, can make the initial management as well as the transition from ductal-dependent to right ventricle-dependent pulmonary circulation challenging; therefore, the optimal timing for intervention and ductal closure needs to be individualized to minimize morbidity and mortality. Regarding patients with an abnormal pulmonary valve (APV), they had a high rate of mortality, and only 15% achieved biventricular circulation. It is not clear whether the operative deaths in this group were associated with an attempt to biventricular repair or single-ventricle palliation. If the case is the former, it is possible that forcing early biventricular repair in the presence of an APV may be associated with worse outcome than if single-ventricle palliation would have been offered. The presence of pulmonary regurgitation in the APV group obviously precluded the presence of pulmonary atresia; however, it was also associated with worse outcome.

On the basis of these observations, it seems that although it could be challenging to mitigate the deleterious effect of a circular shunt, it may take up to 10 days for the antegrade

FIGURE 1. Neonatal Ebstein chest X-ray.

pulmonary blood flow to become apparent in those patients with suitable anatomy. In those with true pulmonary atresia, the decision is simple as long as robust echocardiographic

information could be obtained from the valve, which might not move. This may not be so simple.

Although the optimal management of neonates with Ebstein anomaly remains challenging, progress will only be made after we acquire a better understanding of the anatomic and physiologic variables that play a role. I look forward to the day when the creation of a prospective multi-center registry could provide more definitive answers and inform the management of this complex group of patients; in the mean time we should thank the authors for this contribution.

Reference

1. Baek JS, Yu JJ, Im YM, Yun TJ. Outcomes of neonatal Ebstein's anomaly without right ventricular forward flow. *J Thorac Cardiovasc Surg.* 2016;152:516-21.