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COMMENTARY

Asymptomatic Ebstein's Anomaly in Children and Adults: Intervene or Observe?

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Comments

Ebstein's Anomaly (EA) is a rare congenital heart disease (CHD) with an incidence of approximately 1 in 20,000. The pathognomonic feature involves apical displacement of the septal and posterior leaflets, resulting in valvular insufficiency and right ventricular (RV) remodeling. Untreated patients exhibit a cumulative mortality rate of up to 25% within the first decade, with heart failure (HF) and arrhythmias constituting the predominant causes of death. Current guidelines suggest that asymptomatic patients with accessory pathways may benefit from prophylactic ablation, though robust evidence specific to EA remains limited [1–3].

In recent years, cone reconstruction has emerged as the preferred technique for EA repair due to its superior anatomical restoration, technical reproducibility, and favorable early-to-midterm outcomes. For children (aged >1 year) and adult patients presenting with fatigue (excluding other causes), reduced objective exercise tolerance (e.g., exercise testing demonstrating suboptimal exercise tolerance), reduced arterial oxygen saturation (cyanosis), and exertional dyspnea, multiple authoritative guidelines strongly recommend tricuspid valve repair or replacement surgery [2–4]. Surgery is recommended for patients with severe tricuspid regurgitation (TR) when serial echocardiography or magnetic resonance imaging (MRI) demonstrates progressive RV dilation or dysfunction, regardless of symptomatic status [2–4]. Surgery can be beneficial in asymptomatic patients when there is severe TR, moderate RV enlargement, and valve anatomy favorable for repair [4].

A meta-analysis further supports early intervention for those with progressive RV dilation, demonstrating a 28% reduction in HF risk. While optimal surgical age remains debated, the 2025 American Association for Thoracic Surgery (AATS) guidelines suggested that surgical intervention at 3 to 5 years of age is reasonable in the presence of severe TR with moderate RV enlargement and anatomically repairable valves [4]. And it also highlights individualized timing based on RV function and valve repairability [4]. The data from Boston Children's Hospital reveal that cone reconstruction performed beyond 18 years of age correlates with increased early-phase mortality, whereas procedures conducted before 4 years are associated with accelerated time to tricuspid valve

reoperation during follow-up [5]. Furthermore, the implementation of annuloplasty in children aged ≥8 years demonstrates a significant reduction in long-term TR risk [5]. Recent Mayo Clinic research demonstrates that postponing cone reconstruction until age 4 in patients with stable RV function and absence of HF or cyanosis allows for reductions in early postoperative complications and decreased requirements for Glenn procedures [6].

However, current guidelines lack specific recommendations for asymptomatic children (aged >1 year) and adult EA patients with mild to moderate TR, and the relevant research is also scarce [2–4]. Early intervention may preserve RV function at the cost of accelerated need for tricuspid valve reoperation. Therefore, the decision regarding the timing of surgery requires careful weighing between the durability of tricuspid valve function after repair and the long-term risks of progressive RV dilation and functional deterioration due to delayed intervention. Current evidence demonstrates that cone reconstruction achieves favorable long-term outcomes by effectively restoring tricuspid valve function and attenuating RV remodeling, thus advocating for early intervention to preserve RV functional reserve. While cone reconstruction is the most established approach, modified Danielson techniques have also yielded comparable midterm outcomes in adults at specialized centers. Surgical repair should be performed by congenital heart surgeons with specific expertise in EA procedures [3,4]. Emerging clinical evidence identifies critical prognostic indicators that may necessitate early surgical intervention, including biomarker elevations (B-type natriuretic peptide (BNP) > 100 ng/L, NT-proBNP > 300 ng/L), elevated hemoglobin (Hb)/hematocrit (Hct) levels beyond physiological ranges, cardiac magnetic resonance-derived right-to-left ventricular volume index ratio exceeding 2.5, and Carpentier type C/D morphological profiles demonstrating severe leaflet displacement and annular dilation [7]. These objective parameters should prompt consideration of proactive surgical management in affected individuals, even in the asymptomatic status (NYHA class I) or equivocal symptoms (modified EA functional class 1A).

We advocate that authoritative societies, including the American Heart Association/American College of Cardiology (AHA/ACC) and European Society of Cardiology (ESC), incorporate multimodal parameters—specifically age, imaging parameters, hemodynamic criteria, biomarkers, and anatomical classifications—into intervention criteria for asymptomatic patient management. Similar to the EuroSCORE II model for cardiac surgery risk assessment, an EA-specific online calculator integrating multimodal imaging parameters, biomarker profiles could provide decision support for physicians and families. In the future, multicenter prospective cohort studies are needed to validate the predictive efficacy of the EA-specific online calculator on surgical timing and outcomes. Additionally, through multidisciplinary team collaboration, surgical risks and long-term benefits can be carefully weighed to achieve personalized treatment goals.

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