Ebstein Anomaly

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Risk

- Incidence: Rare; <1:200,000 live births
- Accounts for <1% of congenital heart disease

Perioperative Risks

- Arrhythmias (approximately 20–25% incidence of accessory pathways)
- Intracardiac shunting
- Cyanosis and associated problems (e.g., polycythemia, hyperviscosity, altered vascular function, impaired cardiopulmonary performance)
- Ventricular volume overload
- Ventricular dysfunction

Worry About

- Delayed onset of action of IV agents due to blood pooling into dilated right-sided cardiac structures
- Rhythm disturbances: Atrial and ventricular
- Conditions that may allow for or enhance atrial level R-to-L shunting and increased hypoxemia
- Potential for paradoxical embolism
- Coexisting cardiac defects
- Increases in pulmonary vascular resistance
- Sequelae related to prior cardiovascular interventions

Overview

- Most common form of congenital TV disease.
- Primary congenital cause of TR.
- Results from incomplete delamination of the TV leaflets from the ventricular muscle during cardiac development.
- Wide morphologic spectrum and broad range of anatomic severity. Characterized by (1) downward displacement of septal and posterior TV leaflets attachments toward cardiac apex; (2) adhesion of leaflets to underlying RV myocardium; (3) redundancy of anterior leaflet ("sail-like"); and (4) dilation and thinning of "atrialized" portion of the RV with size reduction of functional chamber (RV myopathy).
- Pathophysiology: Primarily related to degree of TR, extent of RV contractile impairment and reduced compliance, and associated cardiac defects.
- Pts may present at any age.
- Extremely variable clinical spectrum ranging from no-to-minimal symptomatology, to intractable congestive heart failure; in the fetus it can be associated with marked cardiomegaly, arrhythmias, and hydrops.
- Interatrial communication (atrial septal defect or patent foramen ovale) frequently present, resulting in resting or exertional cyanosis due to bidirectional or R-to-L shunting.
- Associated pathology: Pulmonary stenosis or atresia (structural or functional), hypoplastic branch pulmonary arteries, RV outflow obstruction due to abnormal leaflets/chordal structures, ventricular septal defect, patent ductus arteriosus, abnormalities of the mitral and aortic valves, LV noncompaction, LV outflow tract obstruction, coarctation of the aorta.
- Ebsteinoid abnormality of the TV can be seen in association with congenitally corrected transposition of the great arteries.

Etiology

- Heterogeneous: Various genetic, pregnancy, and environmental risk factors implicated.
- Historic association with maternal lithium therapy during pregnancy.
- Most cases are sporadic with rare familial association.

Usual Treatment

- Highly dependent on clinical presentation, physiology, and coexisting cardiac malformations.
- Neonates with heart failure (secondary to severe TR and/or arrhythmias) or cyanosis (due to elevated pulmonary vascular resistance with an associated atrial level R-to-L shunt) may benefit from mechanical ventilation, diuretics, heart failure medications, inotropes, antiarrhythmics, alprostadil (PGE_1), oxygen/iNO, or other pulmonary vasodilators.
- Older children and adults with cyanosis, exertional dyspnea/decreased exercise tolerance due to progressive heart failure, or recurrent arrhythmias are managed with diuretics, antiarrhythmics, and heart failure medications.
- Cath or surgical ablation of atrial arrhythmias, maze procedure, pacemaker for AV block may be indicated.
- Surgery, when necessary, consists of TV repair with annuloplasty or valve replacement, plication of the atrialized RV, and closure of intracardiac communications. If a biventricular repair is not feasible, options include RV exclusion (Starnes procedure), "one and a half" ventricle repair (bidirectional Glenn connection to reduce venous volume load to the RV), or univentricular palliation.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
RESP	Pulm hypoplasia (neonate) Pulm Htn (neonate)	Cyanosis Hypoxia	Tachypnea Resp failure, O_2 sat <90%	CXR, CT scan ABG/VBG, ECHO
CV	RV and/or LV dysfunction Arrhythmias R-to-L shunt	Dyspnea Palpitations Cyanosis at rest or exertion	Wide split S_2, S_3, murmur Tachycardia O_2 sat <90%	CXR, ECHO, MRI, cath ECG, Holter, EP study ABG/VBG, ECHO, cath, exercise test
GI	Liver congestion	Abdominal pain, edema	Ascites, jaundice, hepatomegaly, splenomegaly	Ultrasound/MRI/CT, LFTs (with INR), CBC, EGD
CNS	Paradoxical emboli Stroke	Headache Lethargy, seizures	Neurologic deficits	MRI or CT scan EEG
HEME	Polycythemia Iron deficiency	Headache, dizziness Fatigue	Clubbing Pallor	CBC Peripheral blood smear, ferritin, TIBC

Key References: Dearani JA, Mora BN, Nelson TJ, et al.: Ebstein anomaly review: what's now, what's next? *Expert Rev Cardiovasc Ther* 13(10):1101–1109, 2015; Ross FJ, Latham GJ, Richards M, et al.: Perioperative and anesthetic considerations in Ebstein's anomaly, *Semin Cardiothorac Vasc Anesth* 20(1):82–92, 2016.

Perioperative Implications

Preoperative Preparation

- Appropriate antibiotics for endocarditis prophylaxis.
- Large-bore peripheral IV access as required for planned procedure.
- Available emergency and antiarrhythmic drugs.
- Immediate access to cardioverter/defibrillator.
- Meticulous deairing of IV lines; consider the use of filters.

Monitoring

- Arterial line, CVP, PA cath as indicated; PA cath placement can be difficult and cardiac output measurements erratic.
- Close attention to the ECG.
- Careful monitoring of end tidal CO_2/pulse oximetry/blood gases.
- Urine output to assess renal perfusion.
- Consider the use of transesophageal echocardiographic monitoring in selected cases.

Airway

- Management based on the nature of the procedure
- Potential for bleeding of mucosal surfaces during instrumentation related to increased vascularity in chronic cyanosis

Preinduction/Induction

- Light premedication.
- Consider effects of anesthetic agents/mechanical ventilation on myocardial performance and hemodynamics.
- Potential hemodynamic compromise and rapid deterioration related to hypovolemia (diuretic/vasodilator therapy), myocardial depression, rhythm disturbances.
- Treat reductions in BP and rhythm problems as dictated by the hemodynamics.
- Consider risks and benefits of central neuraxial blockade as appropriate for the procedure.

Maintenance

- Maintain RV preload to provide adequate forward flow.
- Judicious use of fluids in the presence of ventricular dysfunction.
- Maintain a low pulmonary vascular tone to minimize RV afterload and enhance output.
- Normal to high heart rates to sustain forward flow and limit peripheral vascular congestion.
- Consider potential detrimental effects of the surgical procedure on preload/afterload leading to reductions in cardiac output (e.g., increased intraabdominal pressure associated with pneumoperitoneum during laparoscopic procedures).
- Optimize RV and LV function (inotropic support and vasoactive drugs as needed).

Extubation

- Pts may require postoperative mechanical ventilation.

Postoperative Period

- Close monitoring of volume status, UOP/perfusion especially in the setting of baseline ventricular dysfunction
- ECG monitoring for arrhythmias
- Adequate pain control

Anticipated Problems/Concerns

- Potential need for postprocedure/surgery ICU care
- Clinical presentation early in life associated with poor prognosis