20 Tricuspid Atresia

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Tricuspid atresia is a type of single-ventricle CHD that is characterized by the absence of the tricuspid valve, such that there is no direct communication between the RA and the RV. An ASD with an obligate right-to-left shunt is present in all cases and is crucial for survival. Other cardiac anomalies are typically present such as a VSD, transposition of the great arteries, pulmonary atresia or hypoplasia, aortic stenosis, coarctation of the aorta, PDA and a left SVC. The RV is typically hypoplastic, the degree of which depends on the presence and size of the VSD. Other extracardiac anomalies may be present as well.

Classification

The classification of tricuspid atresia is based on the relationship of the great vessels, the presence and size of a VSD, and the amount of pulmonary blood flow:

- Type I: Normally related great arteries (~70%)
  - Ia: No VSD, pulmonary atresia
  - Ib: Small VSD, pulmonary stenosis
  - Ic: Large VSD, no pulmonary stenosis
- Type II: D-transposition of the great arteries (TGA) (~25%)
  - IIa: VSD, pulmonary atresia
  - IIb: VSD, pulmonary or subpulmonary stenosis
  - IIc: Large VSD, no pulmonary stenosis
- Type III: L-TGA (~5%)
  - IIIa: VSD, pulmonary or subpulmonary stenosis
  - IIIb: Subaortic stenosis

A useful way of remembering the subclassification for types I and II is "a" for atresia (pulmonary atresia), "b" for balanced circulation (pulmonary stenosis), and "c" for overCirculation (no pulmonary stenosis).

Pathophysiology and Clinical Presentation

Infants can present with cyanosis, CHF symptoms, or relatively asymptomatic, depending on the amount of pulmonary blood flow and the presence of systemic outflow obstruction.

Infants with decreased pulmonary blood flow (types Ia and IIa) present with cyanosis within the first 24-48 hours of life when the PDA closes. Severe cyanosis can lead to acidosis and shock. Those with unrestricted pulmonary blood flow (types Ic and IIc) present with signs of CHF when the PVR drops. A balanced circulation may be possible in patients with types Ib and IIb.

An important consideration in patients with tricuspid atresia and TGA is that the aorta arises from the hypoplastic RV and flow into the aortic valve is dependent on blood crossing the VSD to reach the aorta. Determining the size and type of VSD (muscular VSDs tend to decrease in size with time) is crucial to prevent the development of

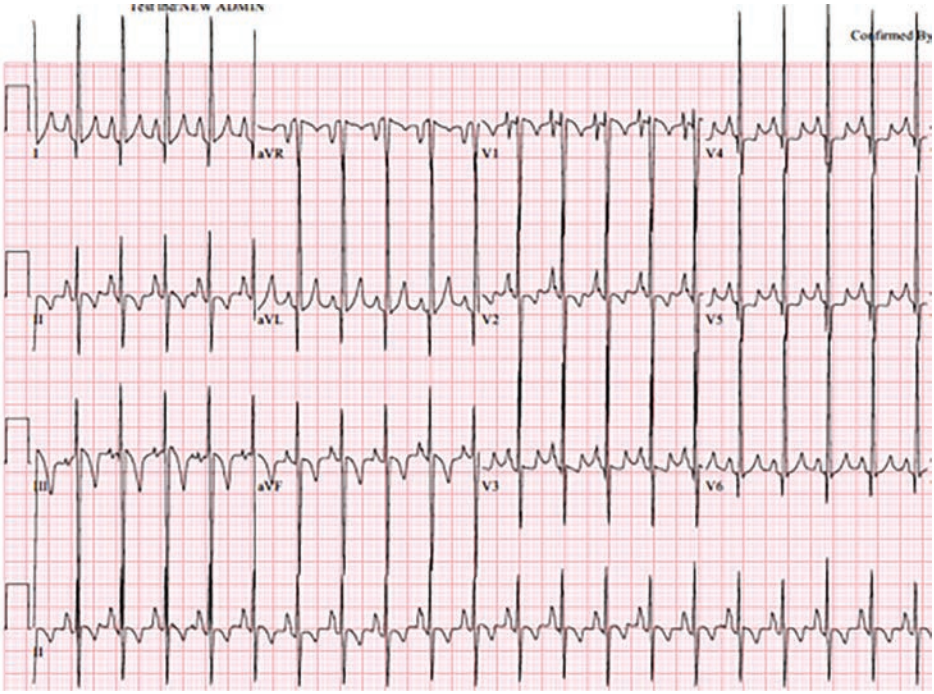

Figure 20-1. ECG on a patient with tricuspid atresia.

systemic outflow obstruction. In general, if a patient with tricuspid atresia and TGA also has an aortic coarctation and/or a hypoplastic aortic arch, it is likely that the VSD is not large enough to sustain the systemic circulation in the long term.

It is important to note that an unrestricted atrial level communication is necessary for survival, and an emergent balloon atrial septostomy may be indicated.

Diagnosis

- **CXR.** The appearance will depend on the amount of pulmonary blood flow. Patients with diminished pulmonary blood flow will have a normal or mildly enlarged cardiac silhouette with decreased pulmonary vascular markings. Patients with increased pulmonary blood flow will have cardiomegaly and pulmonary congestion.
- **ECG (Figure 20-1).** A typical ECG will show RA enlargement, left-axis deviation, and decreased RV forces with LVH.
- **Fetal echocardiogram.** Tricuspid atresia in a fetus is suspected when a hypoplastic RV is seen in a typical 4-chamber view performed routinely. A full study demonstrates the absence of inflow to the RV and a plate-like tricuspid valve. In tricuspid atresia with an intact ventricular septum, the RV and PA will be severely hypoplastic and the PDA will be small. The aorta will typically be enlarged as it carries the combined cardiac output. Other associated lesions will need to be demonstrated clearly, such as the atrial communication, VSD presence and size,

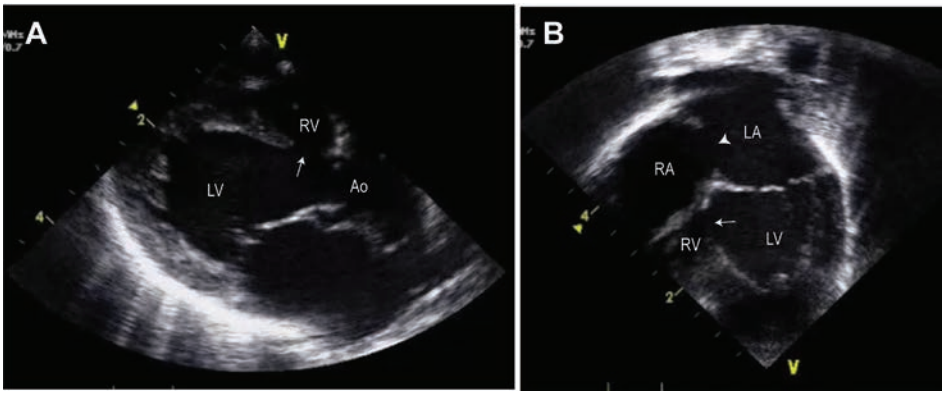

Figure 20-2. Parasternal long-axis (A) and 4-chamber (B) echocardiographic views of a patient with tricuspid atresia and normally related great vessels. There is no communication between the RA and the hypoplastic RV. A large ASD (arrowhead) and a large VSD (arrow) are visualized. Images courtesy of Dr. Josh Kailin, www.pedecho.org.

great vessel relationship, and associated cardiac anomalies. This allows planning for delivery and need for PGE infusion.

- **Postnatal echocardiogram (Figure 20-2).** Echocardiography is the standard imaging technique for diagnosis, showing an absence of the tricuspid valve with a hypoplastic RV. The presence and size of the VSD, the semilunar valves, the relationship of the great vessels, the size of the ASD, and the presence of other cardiac defects need to be well delineated.
- **CTA.** Cross-sectional imaging is typically not required to make a diagnosis of tricuspid atresia. However, it may be useful if the anatomy of the PA, PDA, or aortic arch is unclear on echocardiography.
- **Cardiac catheterization.** Rarely needed for diagnostic purposes. It may be used in certain circumstances for PDA stenting (see Chapter 38).

Surgical Considerations

Patients with tricuspid atresia will require staged surgical palliation towards a Fontan circulation. For details of the single ventricle paradigm, timing of intervention, and surgical techniques, see Chapter 39.

The details of management of patients with tricuspid atresia will depend on the clinical presentation (Figure 20-3). Patients with increased pulmonary blood flow and CHF symptoms will require placement of a PA band (see Chapter 39) in order to protect the pulmonary vasculature prior to the second stage of palliation (bidirectional Glenn). Patients with decreased pulmonary blood flow and significant cyanosis may require administration of PGE to keep the ductus arteriosus open, followed by placement of a shunt or PDA stent (see Chapter 38) in order to provide a stable source of pulmonary blood flow prior to the second stage. Some patients will have a relatively balanced circulation or mild CHF symptoms or cyanosis. These patients can be observed closely until the second stage of palliation, which is usually performed between 4 and 6 months

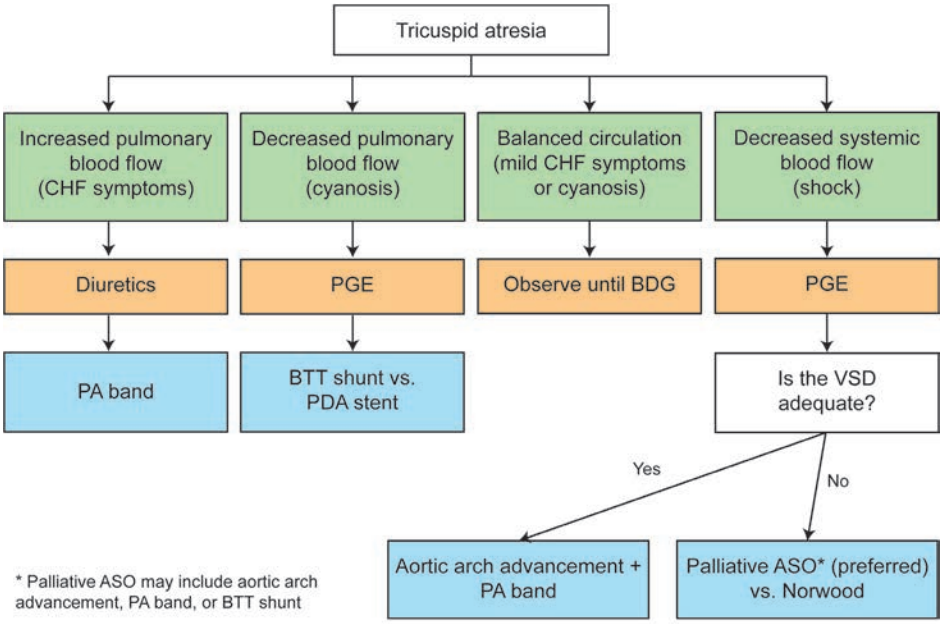

Figure 20-3. Management algorithm for patients with tricuspid atresia based on clinical presentation. ASO: arterial switch operation, BDG: bidirectional Glenn, BTT: Blalock-Taussig-Thomas, PGE: prostaglandin.

of age. However, it is not unusual for patients that start with a balanced circulation to develop significant cyanosis due to progressive closure of the VSD over the first few weeks or months of life.

Patients with tricuspid atresia and TGA pose a challenging situation. A careful assessment is necessary to decide whether the VSD (which serves as the systemic outflow tract) and the aortic arch are adequate to sustain the systemic circulation. If the arch is inadequate and the VSD is considered large enough, the patient may undergo an aortic arch reconstruction (usually an aortic arch advancement, see Chapter 25) and placement of a PA band as a first stage. It is important though not to pursue this approach if the VSD is marginal as the patient will be left with a doubly obstructed heart. A careful determination will then need to be made at the second stage to reassess the size of the VSD. Further narrowing of the VSD may dictate the creation of a Damus-Kaye-Stansel (DKS) anastomosis (anastomosing the proximal ascending aorta with the proximal PA in order to provide unimpeded cardiac outflow) at the time of the Glenn. An incorrect judgment of the VSD at this stage with oversewing of the pulmonary valve and PA at the time of the Glenn may lead to significant subaortic obstruction in the long term, a condition that is challenging to treat and with suboptimal results.

If the VSD (and therefore the systemic outflow) is deemed to be inadequate at the newborn period, there are 2 surgical options: a Norwood-type procedure and a palliative arterial switch operation. Most centers would pursue a Norwood-type strategy

for these patients (see Chapter 27). However, the anteroposterior arrangement of the vessels makes a DKS anastomosis suboptimal and can lead to entrapment of the left PA by the dilated DKS with time. Therefore, the preferred strategy at TCH has been to perform a palliative arterial switch operation in these patients, provided that the coronary anatomy is conducive to such an approach (Heinle et al. 2013). Similar to a traditional arterial switch operation (see Chapter 14), the aorta and PA are switched, including translocation of the coronary arteries. This strategy aligns the LV (with the unobstructed outflow) with the aorta, and the RV (with the obstructed VSD and outflow) with the PA. Most of these patients will also require an aortic arch reconstruction. In these cases, if there is significant discrepancy between the proximal PA and the smaller ascending aorta, the ascending aorta may be enlarged with a small autologous pericardial or homograft patch. If the obstruction at the level of the VSD is not that significant, patients may require, in addition, placement of a PA band in order to further limit the pulmonary blood flow. In some other occasions, placement of an additional Blalock-Taussig-Thomas shunt (see Chapter 38) may be necessary to increase pulmonary blood flow.

Suggested Reading

Heinle JS, Carberry KE, McKenzie ED, et al. Outcomes after the palliative arterial switch operation in neonates with single-ventricle anatomy. Ann Thorac Surg 2013;95:212-218.