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Cardiac CT evaluation requires a systematic approach to evaluate the complex anatomy in congenital heart patients. We suggest the following outline to assess these patients:

1. Systemic and pulmonary veins, atria, and atrioventricular (AV) valves. Start by evaluating the atria and AV valves of the heart and the veins returning blood to the heart. Look for the following:
 - (a) A right superior vena cava (SVC) and inferior vena cava (IVC)
 - (b) A left SVC. Check to determine whether this is solitary or if there is a bilateral SVC with or without a crossing brachiocephalic vein. Also, determine the atrial connection of the SVC; check whether it connects directly to the atrium, or if there is a connection into a coronary sinus.
 - (c) Atrial connection of the IVC. Rarely, it may be bilateral or cross the spine to connect with the contralateral atrium.
 - (d) Pulmonary venous return. Pulmonary veins may return blood anomalously to the systemic circulation, such as the IVC, SVC, or

brachiocephalic vein. Anomalous pulmonary venous return may be partial or total depending on whether all or part of the blood goes to the systemic circulation. It also may be mixed anomalous drainage. Check for a detailed connection of each pulmonary vein.

- (e) Defects in the atrial septum. Types of defects include secundum, primum, patent foramen ovale, and sinus venosus defect. These may be difficult to see and may require viewing different phases of the scan to optimize visualization. It is helpful to correlate these images with echocardiographic examinations.
 - (f) Relative size and appearance of the atria and atrial appendages.
 - (g) AV valve anomalies, including atresia, stenosis, dysplasia (a dysplastic septal leaflet of the tricuspid valve is seen with Ebstein's anomaly), a common AV valve, and thickening of the valves. Valve sizes should be provided. Subtle vegetations may be seen. Correlation with echocardiography may be helpful when subtle findings are suspected.
2. Ventricles
 - (a) Distinguish the morphologic right ventricle (RV) from the left ventricle (LV). The RV typically has a more trabeculated septal surface than the LV; it also has a more pyramidal shape, with a moderator band at its apex. The LV has an ellipsoid shape and a smoother septal surface. The RV outflow tract or infundibulum is muscle bound and typically not in fibrous continuity with the tricuspid valve.

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- (b) Look for ventricular septal defects. These are seen most commonly in the perimembranous region of the ventricular septum but also may be observed in the muscular portion of the septum, subvalvular region, or posteriorly along the septum in patients with AV septal defects.
 - (c) Evaluate ventricular size, motion, and function if sufficient data are available.
 - (d) Look for an abnormally thickened myocardium, which may be a primary or secondary abnormality and may lead to outflow tract obstruction.
3. Great vessels. Assess the following:
- (a) Outflow tracts. They may be switched, narrowed, absent, or aneurysmal.
 - (b) Aorta
 - (i) It may be discontinuous (interruption of the aortic arch), stenotic in patients with coarctation of the aorta or supra-valvular aortic stenosis, or hypoplastic.
 - (ii) Identify whether the arch is on the left or right, and determine the number and location of vessels arising from the arch. Vascular rings, such as a double arch or right arch with an aberrant left subclavian artery, may cause airway obstruction.
 - (iii) Look for aortopulmonary collaterals. These typically are seen along the descending aorta but may arise from the arch or great vessels.
 - (iv) Measure the size and caliber of the aorta at the aortic annulus, sinus of Valsalva, sinotubular junction, transverse arch, and descending aorta. Comparison with normative data (z-score) may be helpful.
 - (c) Pulmonary arteries
 - (i) They may be atretic, hypoplastic, or anomalous. Determine the origin and course of the pulmonary arteries. Some pulmonary arteries may arise from the aorta, patent ductus arteriosus (PDA) or the other pulmonary artery (in pulmonary sling, the left pulmonary artery arises from the right and courses around the trachea, often causing tracheal or bronchial stenosis).
 - (ii) Evaluate the size of the main pulmonary artery and the right and left proximal and distal pulmonary arteries.
 - (iii) Look for other vessels connected to the pulmonary arteries, such as a PDA. Aortopulmonary collaterals may be connected to the pulmonary arteries or may supply the lung directly.
- (d) PDA (patent ductus arteriosus)
- (i) PDA typically originates from the undersurface of the descending aorta or left brachiocephalic/subclavian artery but rarely may have other anomalous origins and may be bilateral, with one part arising from the aorta and the other from the brachiocephalic artery.
 - (ii) It may be large and tortuous, especially in patients with complex congenital heart disease. A diverticulum may be seen at the origin of the ductus (Kommerell's diverticulum). Look for mass effect from the enlarged PDA on other adjacent structures (especially the trachea and bronchi).
 - (iii) Later in life, a calcification often is seen in the region of the ligamentum arteriosum that forms when the ductus closes.
4. Coronary arteries
- (a) Evaluate the origin, number, course, and termination of the coronary arteries. Notify the surgeon of an anomalous course, especially if it crosses an outflow tract.
 - (b) Rarely, a coronary artery may arise from the pulmonary artery, such as in anomalous left coronary artery from the pulmonary artery (ALCAPA).
 - (c) An increased caliber of the coronary arteries in a newborn may suggest a fistulous communication with a low-pressure system, such as the RV or pulmonary artery.
5. Lungs and airways
- (a) Congenital airway anomalies are more common in patients with congenital heart

disease. Congenital stenosis and bilateral left- or right-sidedness is common in patients with asplenia- or polysplenia-type heterotaxies.

- (b) Look specifically for a right upper lobe (pig) bronchus, especially in patients with chronic right upper lobe collapse.
 - (c) Tracheobronchomalacia is common in congenital heart patients. A narrowed horseshoe-like appearance of the airway may be seen on CT.
 - (d) Extrinsic airway compression is very common in patients with complex congenital heart disease. Look for vascular rings, pulmonary sling, and dilated structures compressing the airway.
6. Situs and cardiac position
- (a) Designate each patient as situs solitus (normal), situs inversus (reversed), or situs ambiguous (indeterminate).
 - (b) Situs usually is determined by the sidedness of the liver, stomach, spleen, right atrial appendages, IVC, and cardiac position. In addition, the anatomy of the tracheobronchial tree and the relationship of

the right and left mainstem bronchi with respect to the right and left pulmonary artery are important hallmarks.

- (c) The normal heart is in levocardia (at the left chest), with the apex pointed to the left. Dextrocardia occurs when the heart is in the right side of the chest. In this case, the apex may point to the right or to the left. In mesocardia, the heart is in the midline, with the apex pointed inferiorly, or is difficult to ascertain.
 - (d) If the aforementioned structures are positioned normally, they are in *situs solitus*. If the structures are reversed, they are in *situs inversus*. If there is a combination of both positions, the structures are in *situs ambiguous*.
 - (e) The presence of situs ambiguous may be a hallmark of asplenia and polysplenia heterotaxy syndrome
7. Common surgical procedures in patients with congenital heart disease. When dealing with congenital heart disease, it is vital to understand the basic terminology regarding postoperative shunts, procedures, and surgeries.