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Review

Transthoracic echocardiogram assessment of atrial septal defects in pediatric patients

Vanessa Hormaza[c], Thomas C. Edwards[a,b,*], Heather Mitchell[a], Peace Madueme[a,b], Dima Sawalha-Turpin[a,b], Gul H. Dadlani[a,b]

[a] Nemours Cardiac Center, Nemours Children's Hospital, Orlando, FL, United States of America
[b] Department of Pediatrics, University of Central Florida College of Medicine, Orlando, FL, United States of America
[c] Akron Children's Heart Center, Akron Children's Hospital, Akron, OH, United States of America

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ABSTRACT

Atrial septal defects are the second most common congenital heart defects. Transthoracic echocardiogram is the imaging modality of choice to diagnose atrial septal defects in the pediatric population and delineate the anatomic type of atrial septal defect. This imaging modality not only allows diagnosis but also permits evaluation of concomitant congenital cardiac defects and secondary physiological changes. Clinicians and sonographers must have a thorough understanding of the appropriate echocardiographic views to completely assess an atrial septal defect and neighboring intracardiac structures. This information is essential to determine if a patient is a candidate for a catheterization based intervention or if the patient would be a better candidate for surgical intervention.

1. Introduction

Atrial septal defects (ASD) are interatrial communications that result from a deficiency of the partition between the left and right atrium. They are the second most common congenital heart lesion [1,2]. ASDs comprise 6–10% all congenital heart defects and are the most common form of acyanotic congenital heart disease [3,4]. The worldwide incidence of ASDs is 1.64 per 1000 live births [5]. Patients with an ASD are often asymptomatic for many decades and often times do not present with any clinical findings such as a murmur [6]. Small ASDs may spontaneously resolve, while larger defects tend to grow in size as the patient matures into adulthood [2]. McMahon et al. in 2002 estimated that 65% of large ASDs increased in size over time [2]. Transthoracic echocardiography is the preferred imaging modality for the evaluation of an ASD in children [4]. Echocardiography performed in a systematic approach can easily define the location and anatomic type of the defect. In addition, a clinician or sonographer should utilize the echocardiogram to: define the size and shape of the defect; assess the direction and magnitude of shunting; evaluate secondary physiologic changes to the heart (right heart enlargement, increase in pulmonary blood flow and pulmonary artery pressure); and exclude other forms of associated congenital heart disease [4,7,8] The objective of this article is to review the various types of ASDs and how to thoroughly assess the interatrial septum with transthoracic echocardiography.

2. Embryology

In order to completely understand ASDs, a clinician must have a good knowledge of the embryological development of the interatrial septum. All ASDs can be related to a failure in the normal process of development of the interatrial septum. The septation of the atrium in the human heart is a long and many times incomplete process, seen even into adulthood. From an evolutionary perspective, mammals with placentas have developed a fossa ovalis that allows oxygen rich blood from the placenta to cross the interatrial septum to the left side of the heart and provide the brain and body with higher concentrations of oxygen. This selective advantage allows fetuses to grow larger and have longer gestational periods. The interatrial septum requires three distinct tissues that contribute to the formation of the two-layered interatrial septum: the septum primum, the septum secundum, and the endocardial cushions. Septation starts around week four of gestation, as the septum primum develops and migrates from the superior roof of the atrium. This flimsy first layer of tissue travels inferiorly and fuses with the newly developing endocardial cushions to enclose the ostium

* Corresponding author at: Nemours Cardiac Center, 13535 Nemours Parkway, Orlando, FL 32827, United States of America.
E-mail address: Thomas.edwards@nemours.org (T.C. Edwards).

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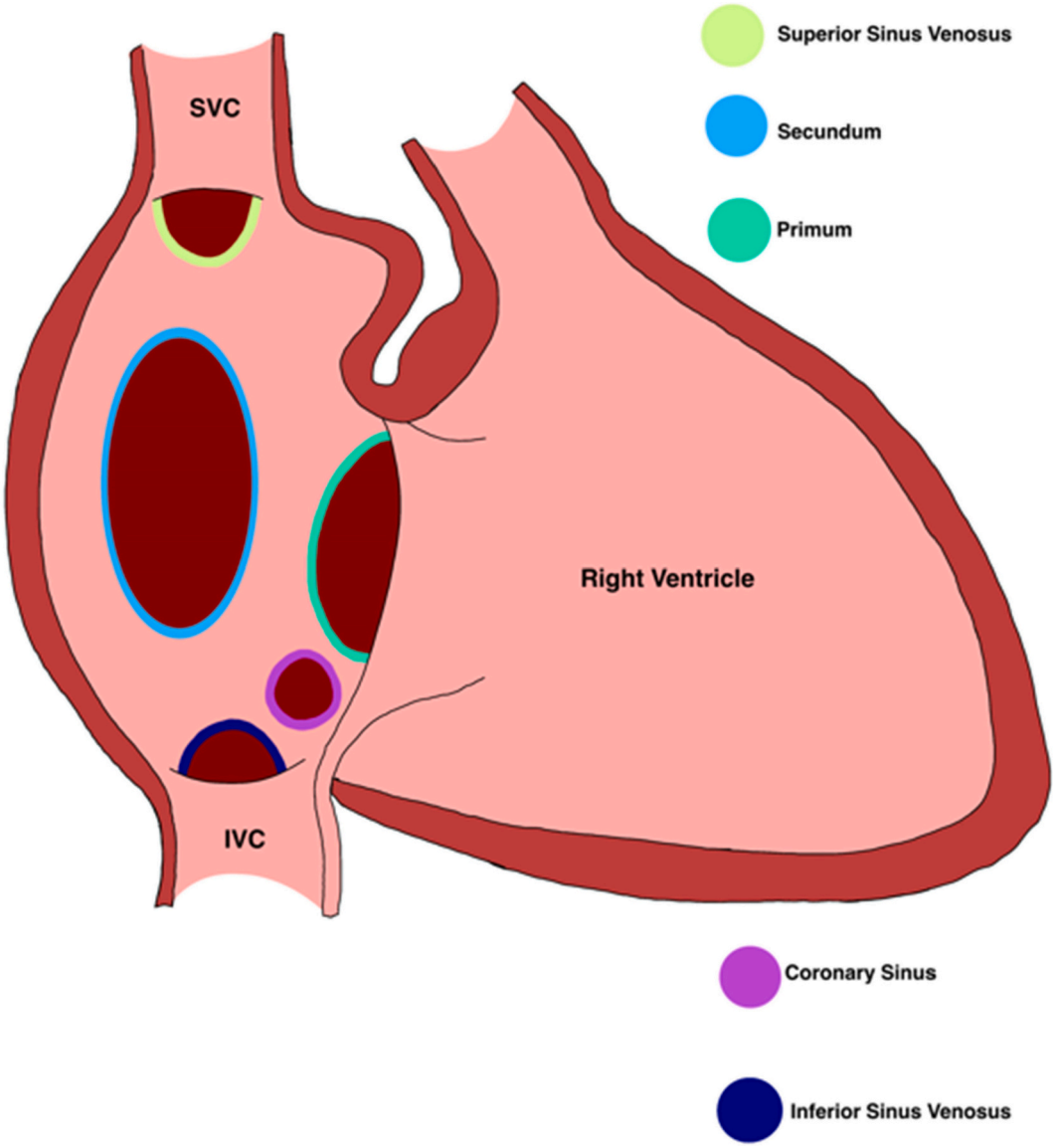


Fig. 1. Types of atrial septal defects.

primum. Failure of the septum primum to attach to the endocardial cushions will lead to a primum ASD (Fig. 1). As the septum primum closes the interatrial septum, small septations start to form in the center of the septum primum and coalesce to form the ostium secundum which occurs during the 5th to 6th week of embryonic life. Simultaneously, the septum secundum (second layer) starts to form as an in folding of the anterior superior atrial roof to the right of the septum primum and grows inferiorly which then partially covers the ostium secundum leaving the foramen ovale [3,4,7–9]. After birth, placental flow ceases and pulmonary venous return from the lungs fills the left atrium and elevates the left atrial pressure, allowing the septum secundum to press against the septum primum and close the fossa ovalis. A patent foramen ovale may persist after birth due to incomplete fusion of the septum secundum to the septum primum (Fig. 1). In addition, secundum ASDs can form if the multiple septations in the septum primum are incompletely covered by the septum secundum (Fig. 1) [3,4,7–9].

The development of the venous system is also important in understanding ASDs. In the primitive heart tube, the sinus venosus has a right and left horn. The right horn of the sinus venosus contributes venous tissue to the inflow of the superior and inferior vena cava as they enter into the right atrium. It is also responsible for separating the right pulmonary vein from the right superior vena cava and the posterior wall of the right atrium. The coronary sinus, which originates from the left horn of the sinus venosus, also becomes incorporated into the posterior smooth wall of the right atrium and is separated from the left atrium by the coronary sinus septum [3,4,7–9]. Developmental failure of the sinus venosus portion of the atrium leads to a superior or inferior sinus venosus type ASD with or without partial anomalous pulmonary venous return (Fig. 1).

2.1. Patent foramen ovale

A patent foramen ovale is not a true atrial septal defect because it does not involve a structural deficiency of the interatrial septum, but is rather a persistent remnant of fetal circulation [4]. The patent foramen ovale is composed of a flap-like valve with the hole being the original fossa ovalis in the center of the septum primum, and the flap being the septum secundum. When a child is born, the patent foramen ovale will have left to right shunting that typically resolves with fusion of the flaps within the first few months of life. In about 20–25% of the population, the foramen ovale remains patent or probe patent [4]. Although a persistent patent foramen ovale is rarely of any clinical concern in

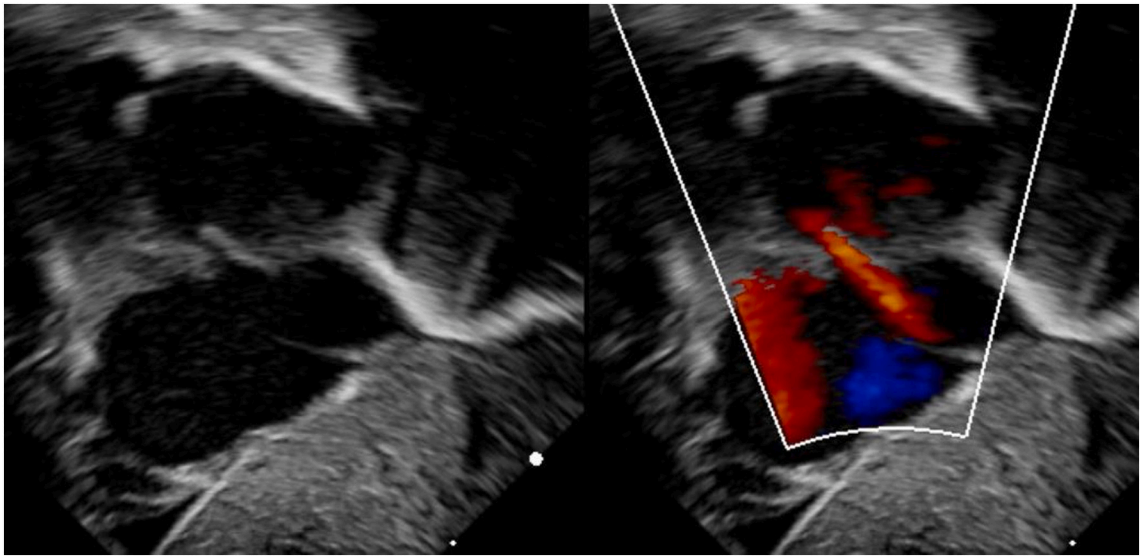


Fig. 2. Patent foramen ovale (PFO).

childhood, it may predispose to paradoxical embolic strokes in hypercoagulable states (Fig. 2).

2.2. Secundum atrial septal defects

A secundum ASD is the most common type of atrial septal defect. Of all ASDs, secundum types account for about 60–80% [3]. A secundum ASD is localized in the central region of the septum where the fossa ovalis is located [4,7]. There are different mechanisms as to how a secundum ASD forms. It can develop when there is excessive absorption of the fenestrations that form in the septum primum and results in patency of the ostium secundum that is unable to be covered by the septum secundum. Other mechanisms include increased flow across the interatrial septum in utero, deficient valve tissues, or deficient growth of the septum secundum [7]. Secundum ASDs are typically elliptical or round in shape and may occur in isolation or in multiples causing a fenestration of the interatrial septum [3,4,7]. The size of ASDs can vary from millimeters to more than 3 cm [4].

An ASD allows for communication between the left and right atria. The amount and direction of blood that crosses the defect depend on the differences in ventricular compliance and the size of the defect, respectively. Typically, the right ventricle is more compliant than the left, resulting in a left to right atrial level shunt. Due to increased blood flow to the right side, progressive enlargement of the right atrium and the right ventricle develops over many years or decades. During this extended period of time, the increased blood flow to the right side may lead to permanent changes within the pulmonary vasculature that can result in the development of pulmonary hypertension [8]. Pulmonary valve stenosis and mitral valve prolapse are commonly associated with secundum ASDs. When evaluating the pulmonary valve in a patient with a co-existing atrial septal defect, clinicians must be mindful that an increased gradient across a pulmonary valve may be secondary to increased flow out of the right ventricular outflow tract, rather than from an intrinsic abnormality of the pulmonary valve [7,8].

2.3. Primum atrial septal defects

Primum ASDs are the second most common and account for about 15–20% of all ASDs [1]. A primum ASD is localized caudally from the area of the fossa ovalis in the anterior-inferior region near the atrioventricular valves (Fig. 3a) [3]. Primum ASDs occur from failure of fusion of the superior and inferior endocardial cushions in conjunction with failure of migration of mesenchymal cells in the septum primum [7]. This lack of fusion results in a primum ASD, but can also cause abnormal development of the atrioventricular valve. Primum ASDs are typically within the spectrum of atrioventricular canal defects and therefore can present in isolation or with a ventricular component forming a canal defect [4,7]. When in isolation a primum ASD is known as a partial atrioventricular canal defect. The atrioventricular valves are usually abnormal in the sense that they consist of one valve annulus with two orifices. The mitral valve in this lesion typically has a cleft with the common complication of mitral regurgitation [4,7,8] (Fig. 3b).

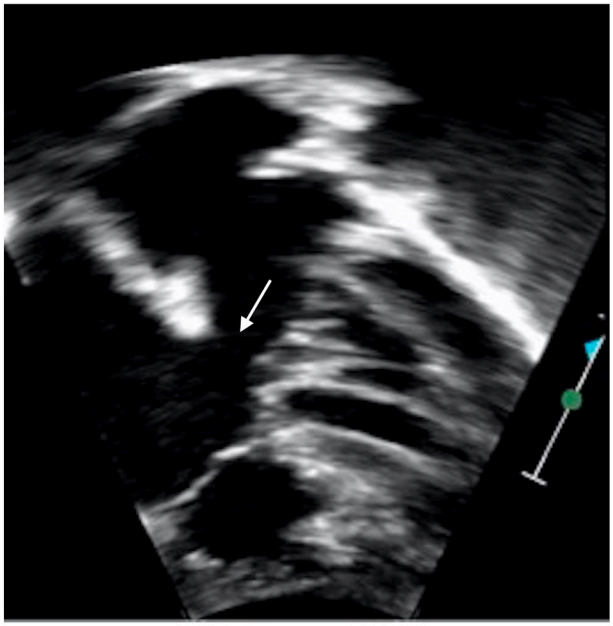
Other associated defects include persistent left superior vena cava to the coronary sinus, parachute mitral valve, and double orifice mitral valve. These defects typically present earlier in life. Electrocardiogram characteristics include the presence of a superior QRS axis [3,7]. A primum ASD is not amenable to percutaneous transcatheter device closure and requires surgical repair due to its location and often the associated cleft in the anterior leaflet of the mitral valve which may need to be addressed at the time of surgery as well [4,7] (Fig. 3b).

2.4. Sinus venosus defects

Sinus venosus defects account for 5–10% of all ASDs and are not true defects of the interatrial septum [1]. Sinus venosus defects result from the partial or entire lack of formation (from excessive reabsorption) of the sinus venosus septum causing a deficiency of the tissue that separates the right pulmonary veins from the right superior vena cava and the posterior wall of the left atrium [3,4,7,8]. This results in the superior vena cava being displaced over both the right and left atrium. The right upper pulmonary vein may then anomalously drain into either the superior vena cava or drain directly into the right atrium (Fig. 4a). This describes a superior sinus venosus ASD. An inferior vena cava sinus venosus ASD, is inferior to the fossa ovalis where there is deficiency of the tissue between the inferior vena cava and the right lower pulmonary veins (Fig. 4b). This is not as common as the superior defect.

A pediatric cardiologist or sonographer should be suspicious of a sinus venosus type ASD where there is not an obvious explanation for enlargement of the right atrium and ventricle. Sinus venosus defects usually warrant further imaging to confirm the diagnosis. This can be in the form of a cardiac computed tomography or cardiac magnetic resonance imaging [[3]] (Fig. 4c). Additional associated abnormalities to sinus venosus defects include a persistent left sided superior vena cava.

a: Primum atrial septal defect



b: Cleft mitral valve

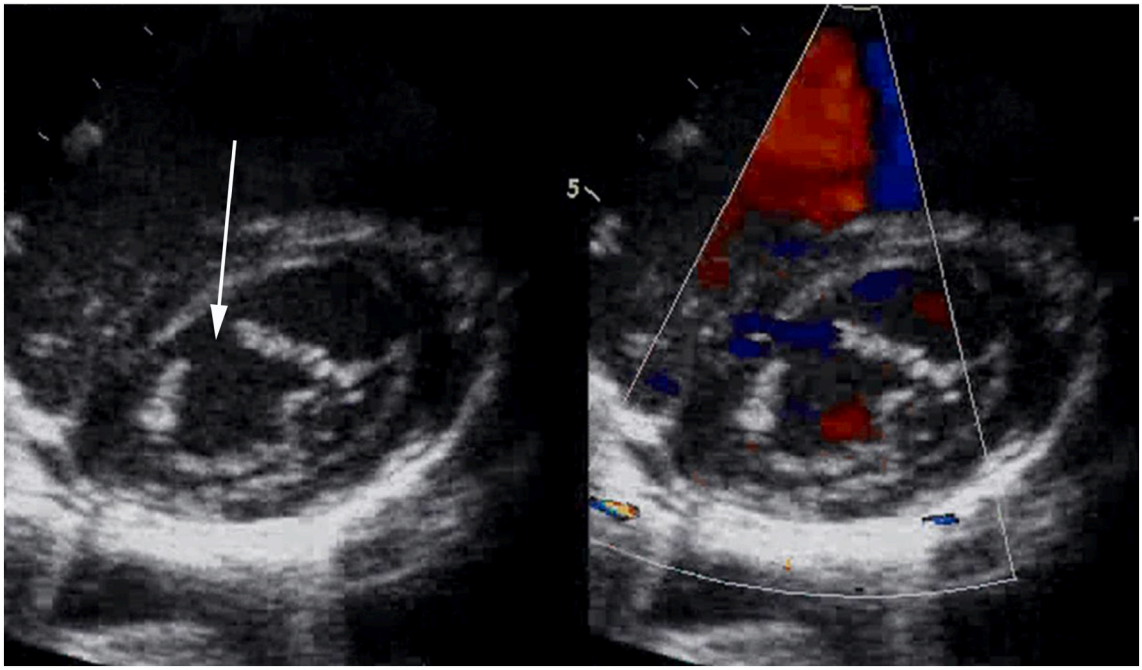


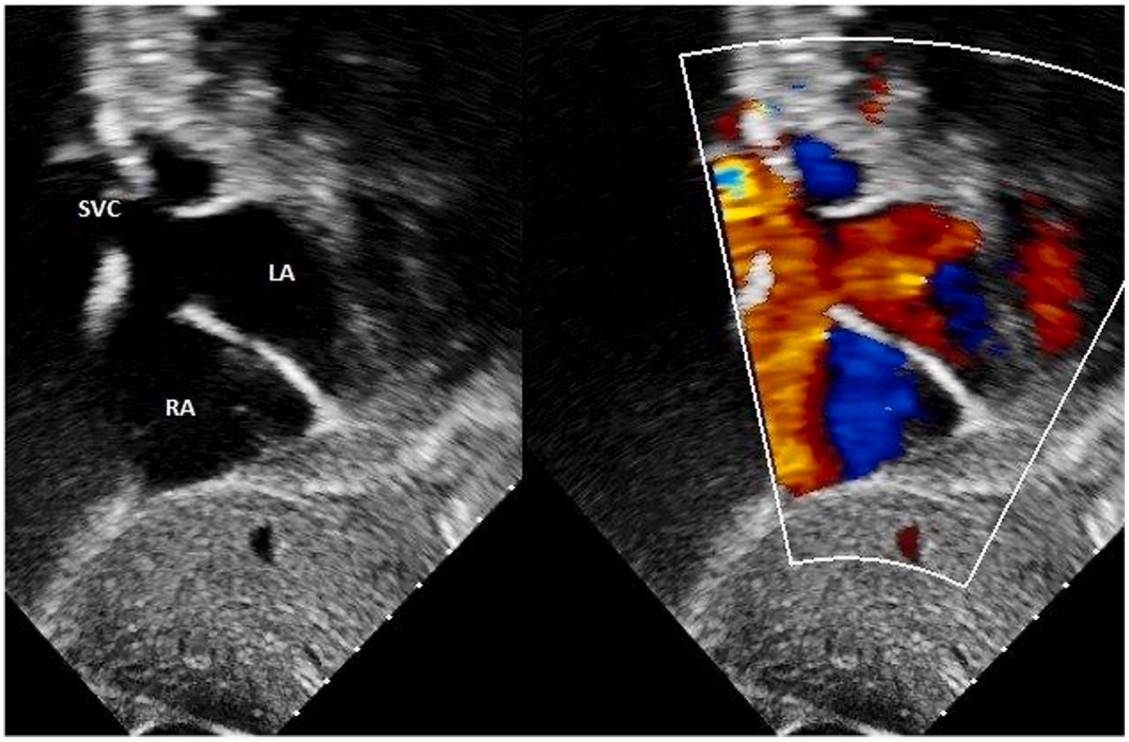
Fig. 3. a: Primum atrial septal defect.
b: Cleft mitral valve.

A sinus venosus type ASD cannot be repaired through transcutaneous closure and requires surgical repair because of the need to baffle the pulmonary veins to the left atrium [[4]].

2.5. Coronary sinus defects

Coronary sinus type defects are extremely rare and when in isolation account for less than 1% of all ASDs. Coronary sinus ASDs occur because of failure of development of the coronary sinus septum which is the tissue that separates the left atrium from the coronary sinus. The defect can either be partial or complete and is often referred to as being unroofed [3,4,7,8]. This results in blood from the left atrium entering through a deficient septum and emptying into the right atrium through the os of the coronary sinus. Coronary sinus defects may occur in association with a persistent left superior vena cava, but may also be present in the absence of a left superior vena cava. Diagnosis of this

a: Sinus venosus atrial septal defect



b: Inferior sinus venosus atrial septal defect

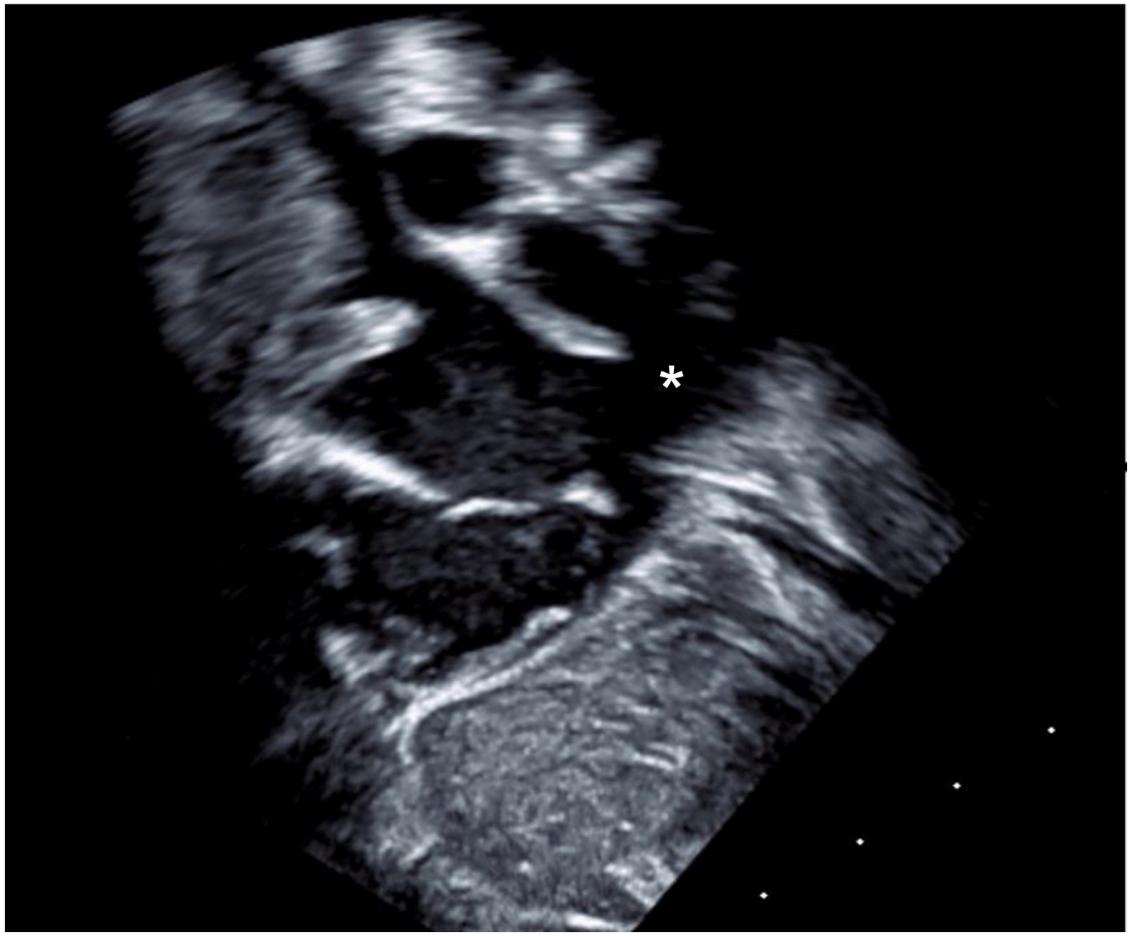


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Fig. 4. a: Sinus venosus atrial septal defect.
b: Inferior sinus venosus atrial septal defect.
c. Magnetic resonance angiography of partial anomalous pulmonary veins associated with a sinus venosus atrial septal defect.

defect may be difficult using transthoracic echocardiography, but this is another lesion that should be taken into consideration when there is unexplained right atrial and ventricular enlargement. To confirm this lesion, an agitated saline test is performed in conjunction with the transthoracic echocardiogram. A positive test would demonstrate bubbles which appear in the left atrium first then in the right atrium when agitated saline is injected into the left arm (Fig. 5). This defect requires surgical repair and is not a candidate for percutaneous transcatheter device closure. Confirmation of a left superior vena cava is vital to surgical planning [3,8].

2.6. Echocardiographic evaluation of atrial septal defects

The echocardiographic evaluation of a secundum ASD requires the utilization of multiple complementary views and planes. The goal of echocardiography is to determine the location, size of the defect, size of the surrounding rims, and distance from other surrounding structures especially the superior and inferior vena cava, pulmonary veins, coronary sinus, and atrioventricular valves. Visualization of hemodynamic consequences such a right atrial and ventricular enlargement, pulmonary artery dilation, right ventricular, and pulmonary artery

c. Magnetic resonance angiography of partial anomalous pulmonary veins associated with a sinus venosus atrial septal defect

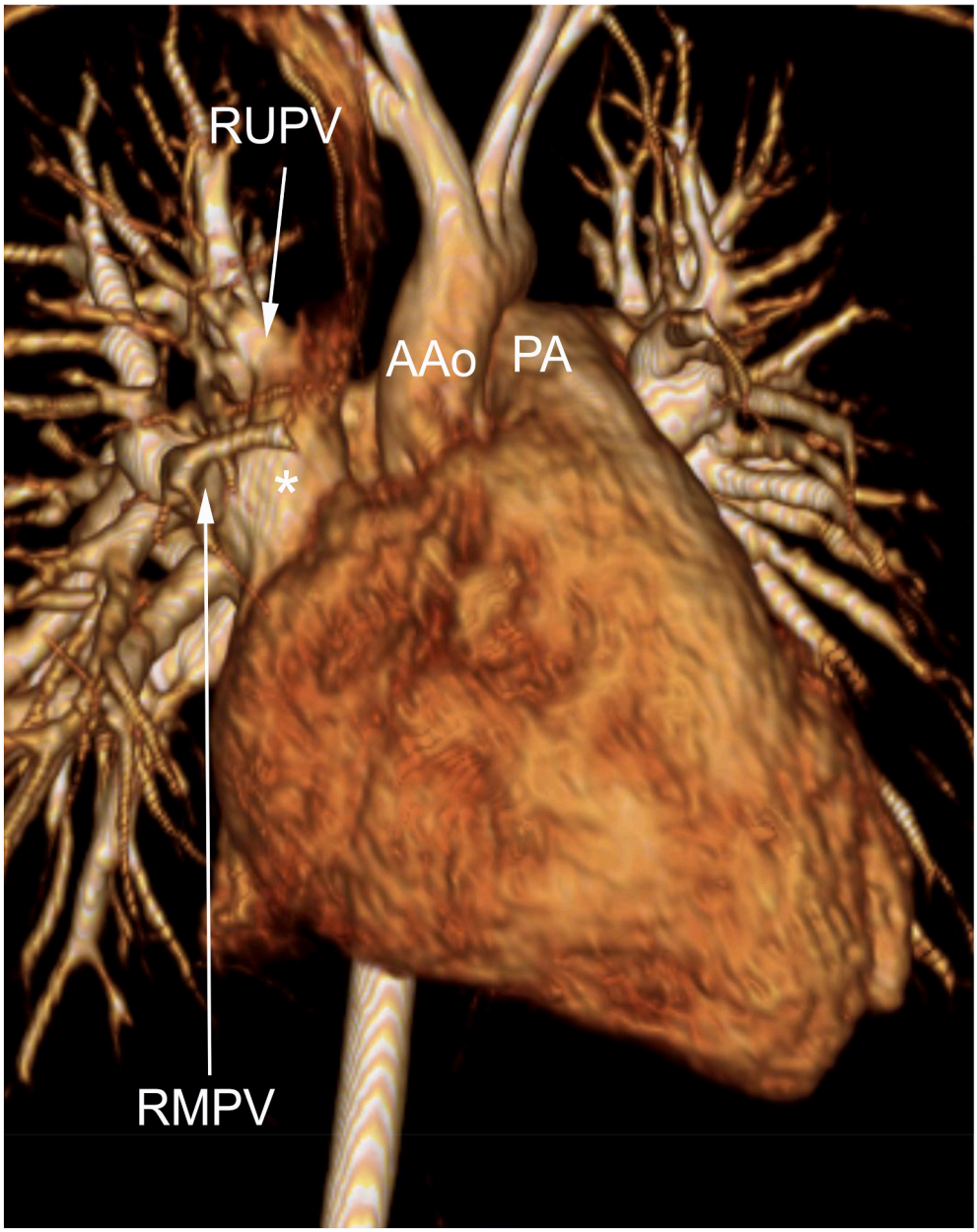

Fig. 4. (*continued*)

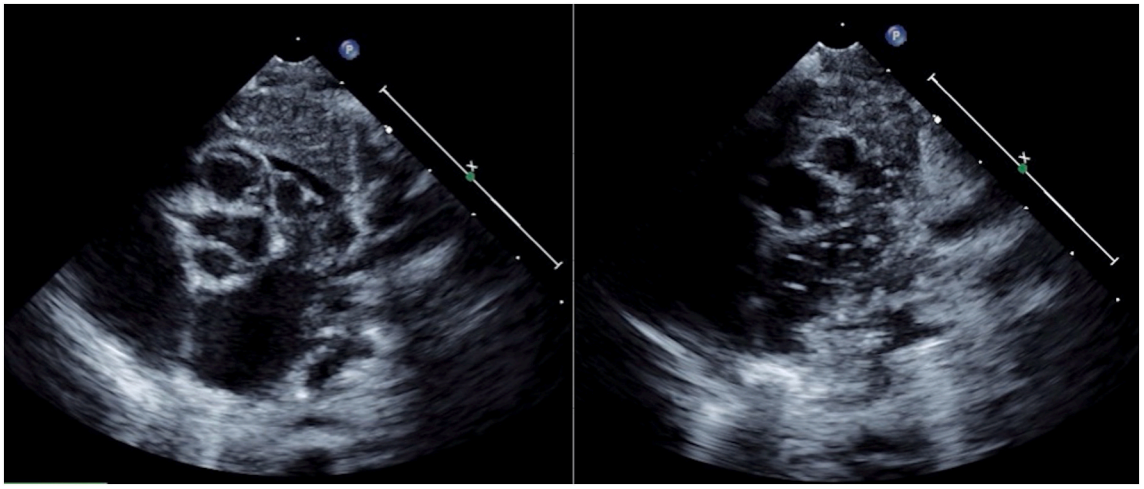

Fig. 5. Coronary sinus atrial septal defect.

pressure elevation can be determined by two dimensional echocardiography in conjunction with spectral and color Doppler. Investigation of associated cardiac abnormalities is necessary to obtain a complete picture of the hemodynamic and clinical state of the patient. The importance of obtaining all information allows for a decision to be made in regards to candidacy for percutaneous transcatheter closure versus primary surgical closure and the timing of repair [4,7,8].

2.7. Subcostal imaging

Subcostal imaging in the pediatric population is the preferred view for evaluation of ASDs because it allows for better visualization of the defect and its surrounding structures. From this view, the interatrial septum lies perpendicular to the plane of the ultrasound beam in both the coronal and sagittal planes. This view prevents any artificial dropout and enables the evaluator to confidently view an ASD when present [4,7,8].

2.8. Subcostal coronal

The subcostal coronal view, or the subcostal four chamber view, shows the interatrial septum along its anterior-to-posterior axis. Sweeps are performed form an anterior to posterior direction. In this view, the relationship of the defect to the superior vena cava and the right pulmonary veins can be seen. Angling the ultrasound probe more anteriorly and superiorly displays the interatrial septum just posterior to the aorta. Angling more posterior allows the posterior-inferior portion of the interatrial septum to come into view. The addition of color Doppler provides information in regards to the direction of blood flow across the ASD (Fig. 6). The direction of flow will help provide information in regards to the clinical state of the patient. The shunt direction is typically from left to right, but in the setting of decreased right ventricular compliance or elevated pulmonary artery pressure, the shunt will be either bidirectional or all right to left. In addition, this view may help better visualize if there is more than one defect or if the septum is fenestrated. Pulse wave Doppler can be performed across the ASD and should show continuous, low velocity phasic flow. Pulse wave Doppler can also demonstrate any gradient between the left and right atria which would alter the Doppler signal resulting in a higher velocity non-phasic flow [4,7,8]. A subcostal coronal view is where a typical secundum ASD is best visualized. It allows for measurement of the ASD along its long axis (Fig. 6).

2.9. Subcostal sagittal

The subcostal sagittal view is obtained by rotating the transducer 90 degrees from the position of the subcostal coronal view (Fig. 6b). From this view, the superior-inferior axis of the interatrial septum is visualized. This is an optimal view to evaluate the anterior-superior and inferior-posterior rims of an ASD. Angling the transducer left and right shows the relationship between the superior vena cava, the rims of the ASD, and the pulmonary veins, in particular the right-sided veins. This view also permits evaluation of the right ventricle, right ventricular outflow tract, pulmonary valve, and pulmonary artery with both two-dimensional echocardiography, spectral Doppler, and color Doppler [4,7,8].

From this view, measurements of the superior-anterior rim of a secundum ASD can be obtained just below the junction of the superior vena cava and the right atrium, in addition to measurement from the defect to the inferior vena cava. The measurement of the defect in this plane in comparison to the long axis allows for determination of the shape of the defect. This is also a good view to see how the right upper pulmonary vein normally connects to the left atrium posterior to this rim, therefore making the subcostal sagittal an optimal view to diagnose a sinus venosus type defect. For a superior sinus venosus defect, there is dropout of the tissue that separates the entrance of the superior vena cava into the right atrium and the right pulmonary vein (Fig. 4) [4,7,8].

2.10. Parasternal long axis

The parasternal long axis view does not allow direct visualization of the interatrial septum. Rather this view allows for collection of information in regards to secondary changes that occur as a result of a significant atrial level shunt. In this view, the right ventricle is visualized and it can be determined if there is enlargement of the right ventricular cavity. The interventricular septum is very well visualized and any abnormal motion, i.e. paradoxical motion of the interventricular septum with flattening in diastole, secondary to a right ventricular volume overload, can be appreciated. The mitral valve is also well demonstrated in this view and the presence of mitral valve prolapse and/or regurgitation can be seen. A dilated coronary sinus (Fig. 7) can also be seen just posterior to the mitral valve in the atrioventricular groove, which could give additional information to support the presence of a persistent left superior vena cava or lead to suspicion of a coronary sinus type ASD.

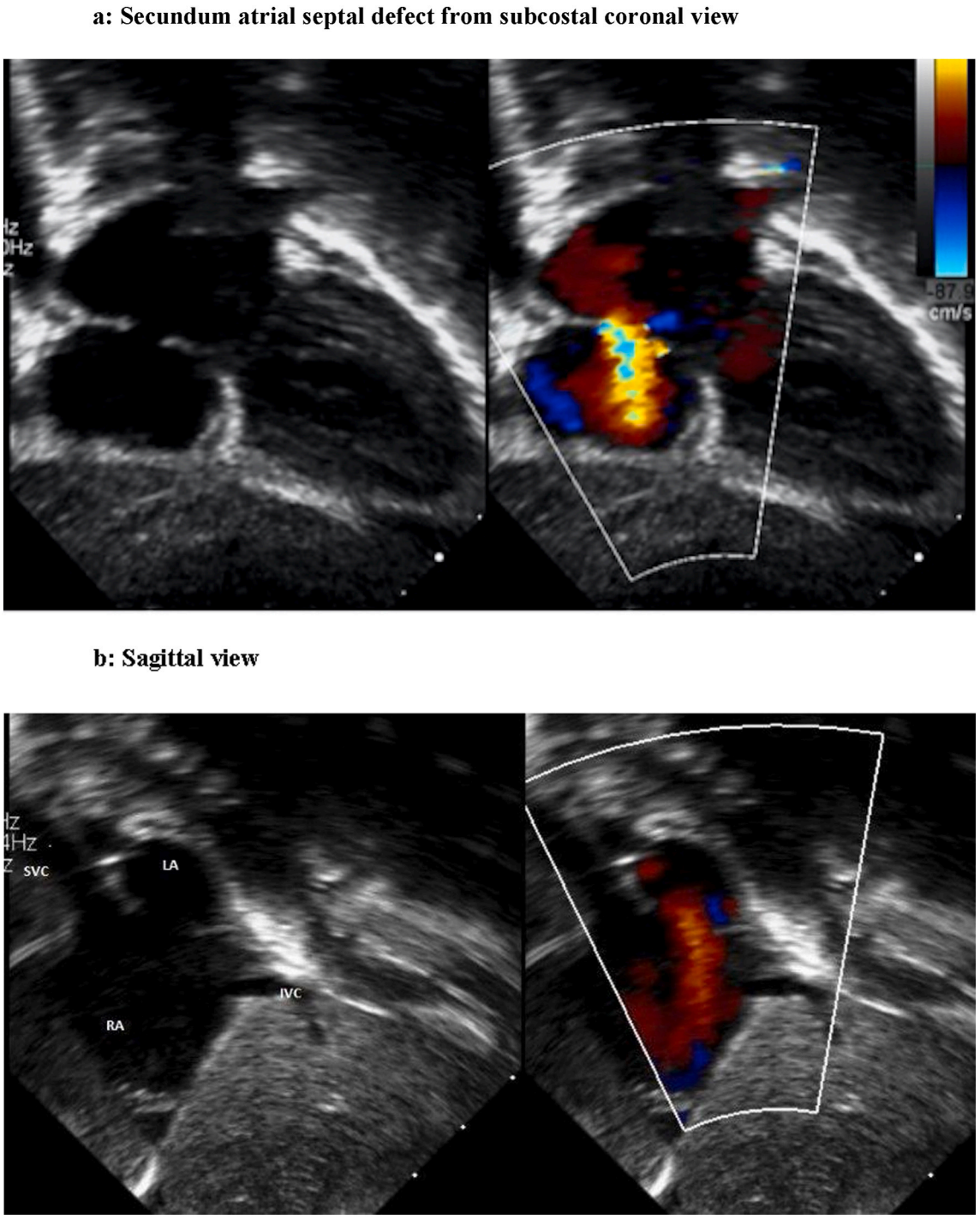

Fig. 6. a: Secundum atrial septal defect from subcostal coronal view
b: Sagittal view.

Angulation of the transducer inferiorly brings the tricuspid valve into view and in conjunction with color and spectral Doppler allows for detection and evaluation of tricuspid regurgitation. The velocity of tricuspid regurgitation allows for the estimation of right ventricular systolic pressure using the modified Bernoulli equation [7]. This allows one to compare right ventricular pressure to systemic pressure to determine if there is pulmonary hypertension [4,7,8].

Superior angulation of the transducer brings the pulmonary valve, right ventricular outflow tract, and main pulmonary artery into view [8]. As previously discussed, pulmonary valve stenosis may be an associated finding and can be well evaluated in this view. The valve leaflet characteristics such as thickening and doming can be evaluated. The velocity across the pulmonary valve can be obtained using spectral Doppler to determine if any stenosis is present. A velocity measurement up to 2.5 m/s or a gradient of 30 mmHg across the pulmonary valve is considered normal in the setting of a hemodynamically significant atrial level shunt. A velocity measurement greater than 2.5 m/s or a gradient greater than 30 mmHg would be consistent with pulmonary valve stenosis [7]. This angulation is also important to determine if there is dilation of the main pulmonary artery segment which can also be present as a secondary outcome of a significant or long standing atrial level shunt [4,7,8].

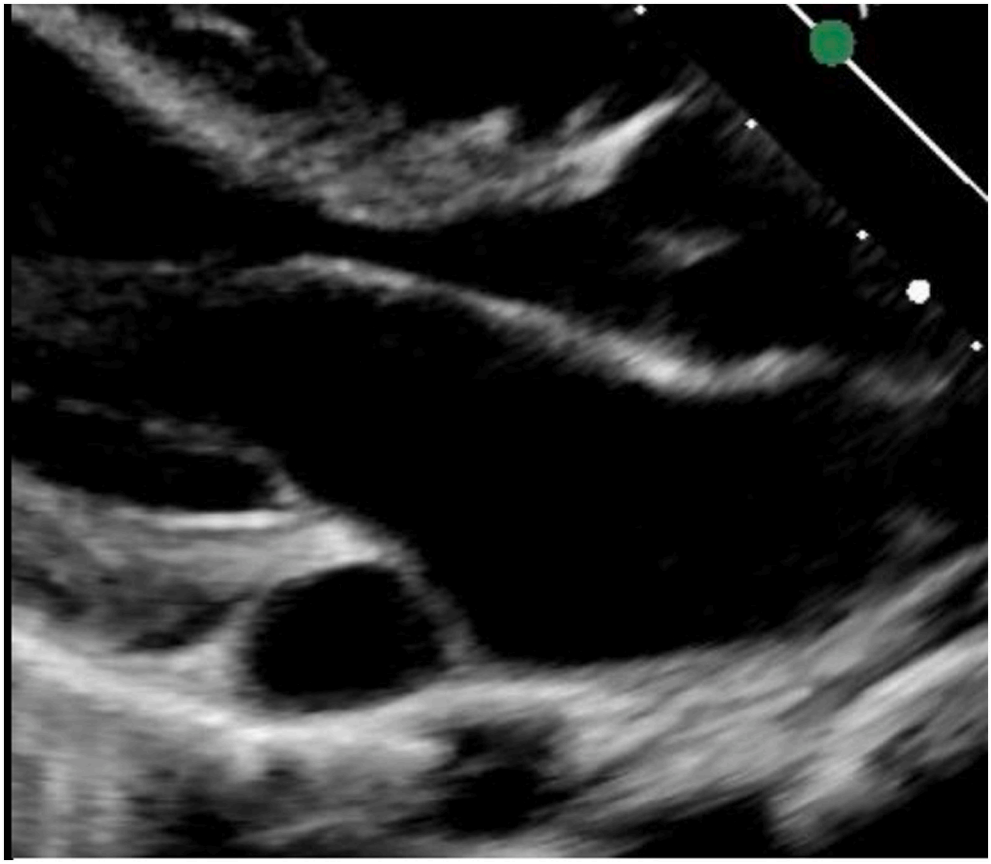

Fig. 7. Parasternal long axis - coronary sinus (CS) dilation.

2.11. Parasternal short axis

The parasternal short axis view allows visualization of the retro-aortic region of the atrial septum, which is important for evaluation prior to consideration of percutaneous transcatheter intervention (Fig. 8). The transducer ultrasound beams are perpendicular to the interatrial septum in this view and visualization of the shunt direction is useful with color Doppler. Sinus venosus type defects and posterior inferior secundum atrial septal defects are easily seen from this view. The right atrium, tricuspid valve, and right ventricle are also well visualized. Additionally, any enlargement of right sided structures, septal flattening, and tricuspid regurgitation can be seen in this view [4,8].

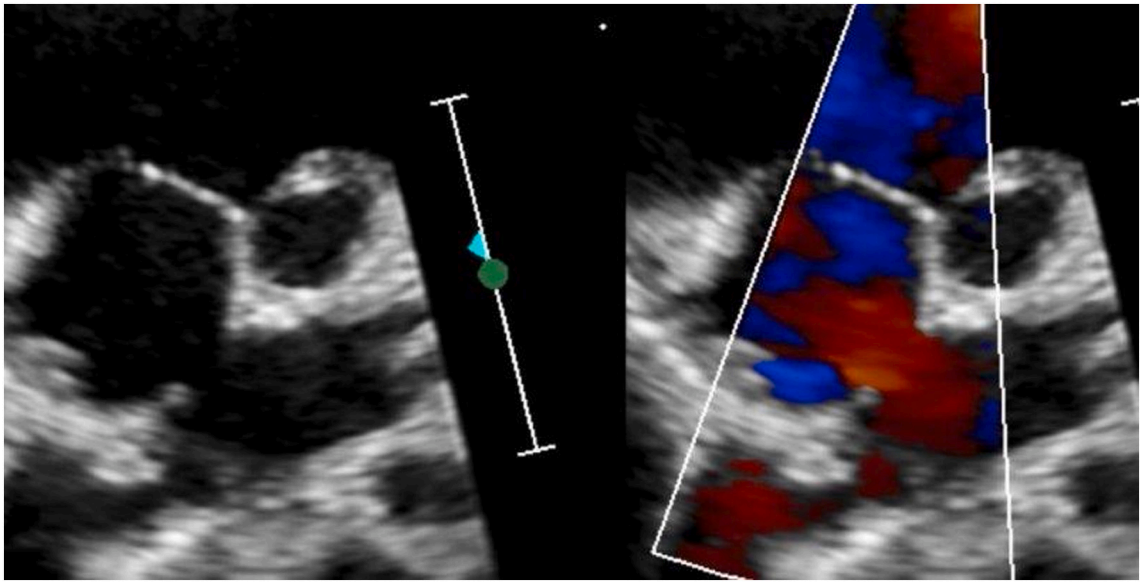

Fig. 8. Parasternal short axis view – showing retro-aortic rim in a secundum atrial septal defect.

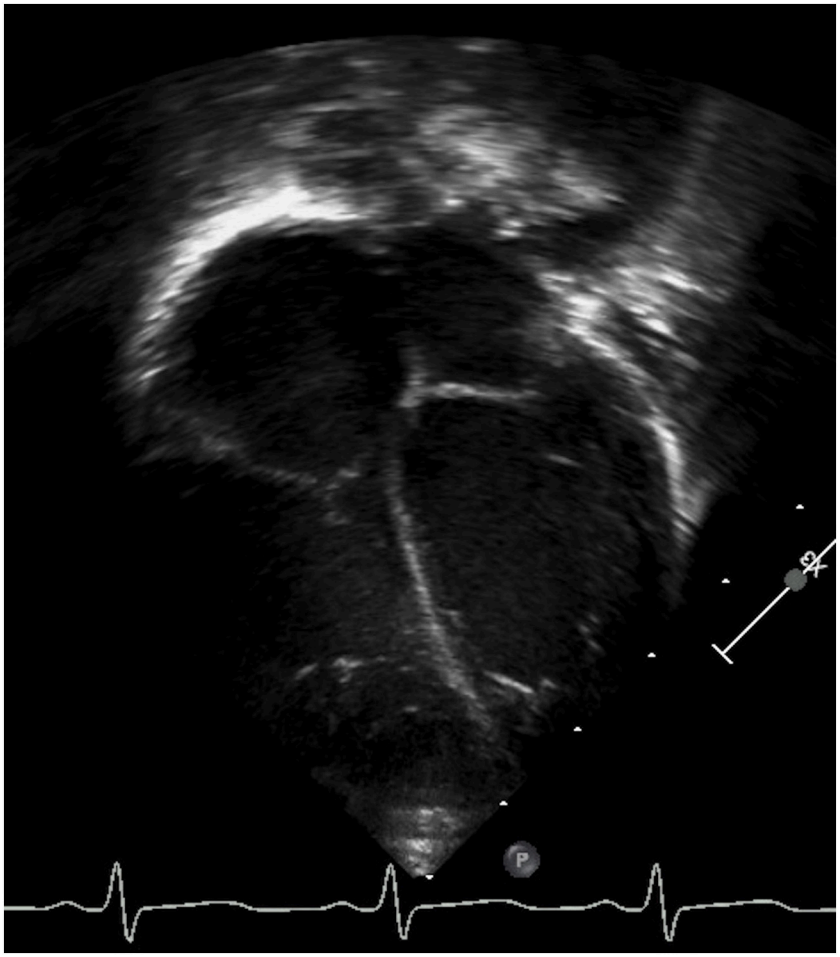

Fig. 9. Apical four chamber view with right atrial and right ventricular dilation.

2.12. Apical four chamber view

The apical four chamber view is best utilized to visual secondary changes that occur with a hemodynamically significant atrial level shunt. From this view, one can qualitatively and quantitatively evaluate for right atrial and right ventricular enlargement (Fig. 9). This view allows for another opportunity to evaluate for tricuspid regurgitation and estimation of right ventricular systolic pressures. Evaluation of the mitral valve for any abnormalities or the presence of mitral stenosis should be a part of the investigation as this could further exacerbate an atrial level shunt if present. The apical four chamber view is not a reliable view to visualize an ASD because the septal tissue in this view is parallel to the ultrasound beam from the transducer which can produce artifact drop out of the interatrial septum, creating an illusion of an ASD, or leading to an overestimation of the size of an existing ASD. This view, therefore, should be used in conjunction with other views that better visualize the interatrial septum. Anterior angulation will bring the main pulmonary artery and right ventricular outflow tract into view, allowing for another opportunity to measure the pressure gradients across these structures using spectral Doppler [4,7,8].

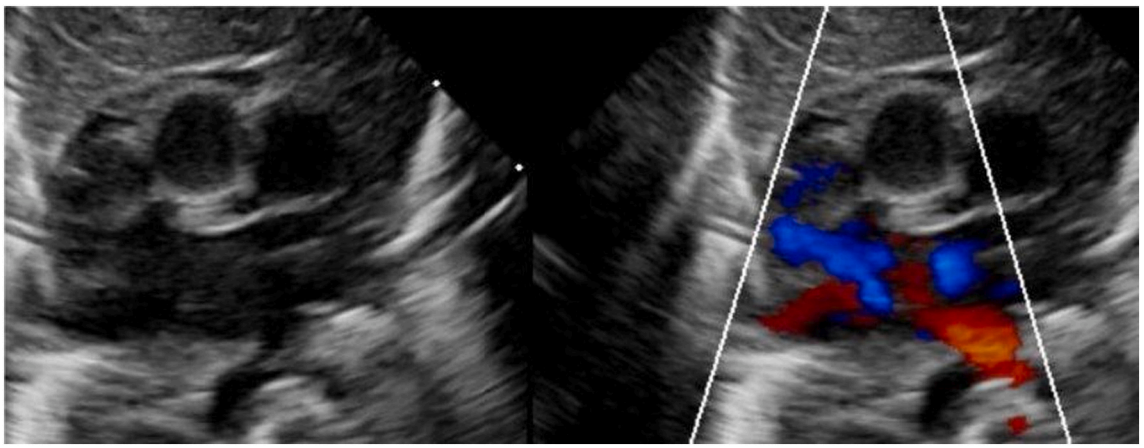

Fig. 10. Suprasternal notch. Crab view showing pulmonary venous return.

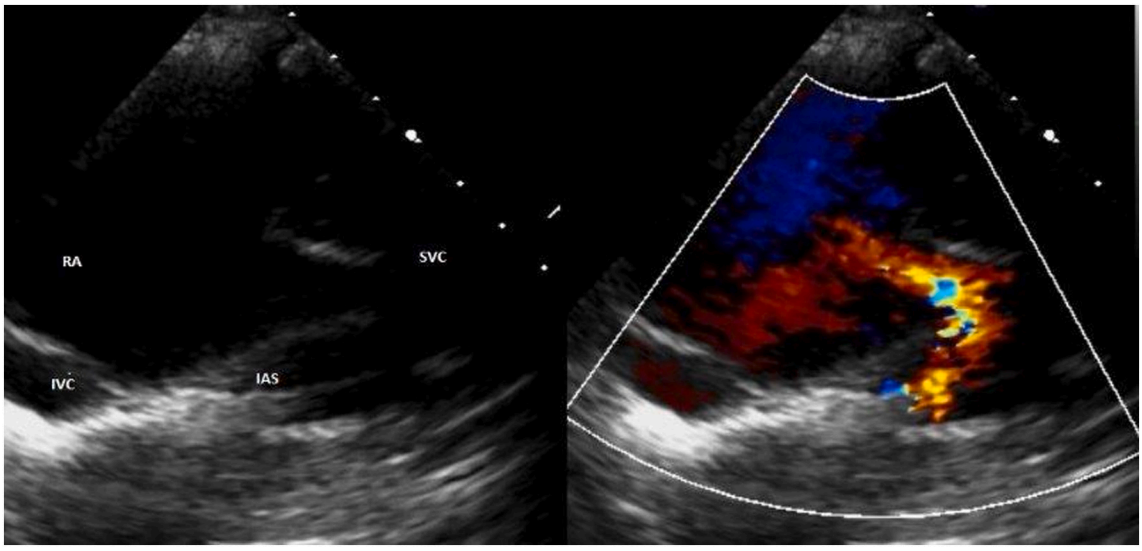

Fig. 11. High right parasternal view of atrial septum – sinus venosus atrial septal defect.

2.13. Suprasternal notch view

From the suprasternal notch view, the pulmonary veins can be seen returning to the left atrium, which is the more inferior structure. This view is also known as the crab view and also enables the visualization of the right and left pulmonary arteries (Fig. 10). Rotation from this view to the sagittal view can reveal an anomalous left pulmonary vein draining into a vertical vein. The color Doppler in this case will demonstrate flow in the direction of the transducer in a superior fashion, draining into the vertical vein [8].

2.14. High right parasternal view

The high right parasternal view may be especially helpful in the diagnosis of a sinus venosus defect, especially if the subcostal views are suboptimal. This view is obtained with the patient in the right lateral decubitus position [4]. With the probe placed in a superior-inferior plane, the superior (cephalad) aspect of the interatrial septum and the junction between the superior vena cava and the right atrium, can be visualized [8]. An inferior sinus venosus ASD may also be best visualized here as the superior and inferior portions of the atrial septum can be visualized (Fig. 11) [4,8].

3. Conclusion

The interatrial septum develops from a complex embryological process that may lead to anomalies in multiple locations. Extensive knowledge of the atrial anatomy and embryology is required for clinicians and sonographers who perform or interpret echocardiograms in the pediatric population. A thorough and systematic echocardiogram is necessary in order to completely assess the interatrial septum. The information gathered through echocardiographic images allows a clinician to diagnose ASDs, determine therapeutic options, and longitudinally manage patients with congenital anomalies of the interatrial septum.

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Declaration of competing interest

Authors have no conflicts of interest to declare.

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