

Case Report

A Case Report: Atrioventricular Block Secondary to Ruptured Sinus of Valsalva Aneurysm in A 12-Year-Old

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Abstract:

Ruptured aortic sinus of Valsalva aneurysm is a rare cardiac anomaly that can lead to life-threatening complications. We present a case of a 12-year-old girl with complete heart block, who presented with these symptoms and was diagnosed with a ruptured aortic sinus of Valsalva aneurysm dissecting into the interventricular septum, with probable compression of the conduction system. Echocardiography and AngioTDM confirmed the diagnosis, and the anomaly was successfully managed percutaneously by closing the orifice of communication using a ventricular septal defect (VSD) muscular occluder. Follow-up examinations showed no flow inside the aneurysm, indicating a successful closure procedure. This case highlights the importance of early diagnosis and prompt intervention in managing ruptured aortic sinus of Valsalva aneurysm in pediatric patients.

Keywords: Atrioventricular block, Sinus of Valsalva aneurysm, Interventricular septum, Percutaneous intervention, Muscular VSD occluder.

Introduction:

Ruptured aortic sinus of Valsalva aneurysm is a rare cardiac anomaly that can have severe consequences, including cardiac arrhythmias, heart failure, and even sudden death. The condition is particularly challenging to manage in pediatric patients due to their smaller anatomy and higher risk of complications. We present a case of a 12-year-old girl who presented with a complete atrioventricular block, syncope, due to ruptured aortic sinus of Valsalva aneurysm dissecting into the interventricular septum, successfully managed percutaneously.



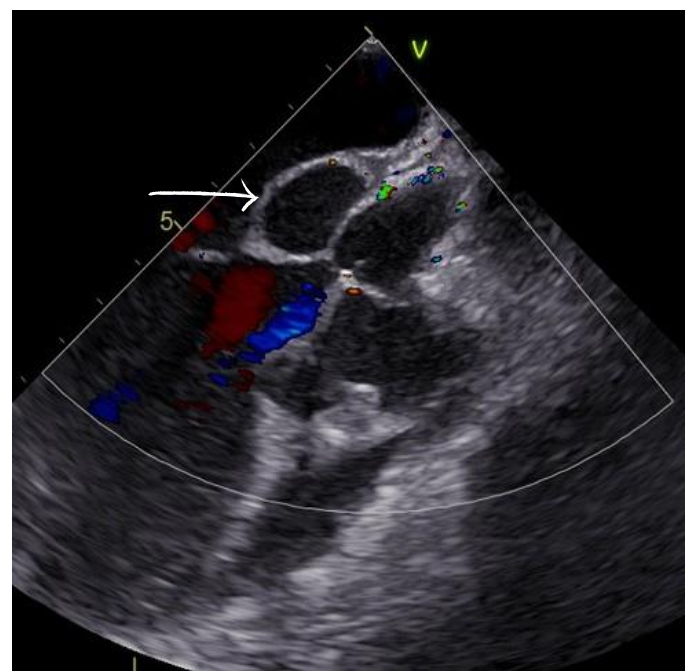
4 chambers view Dissection of the interventricular septum

Case Presentation:

A 12-year-old girl presented with episodes of syncope and a cardiac rate of 45 beats per minute. The EKG found a complete heart block with enlarged QRS, heart rate of 45 B/M

Diagnostic Assessment:

Echocardiography revealed:



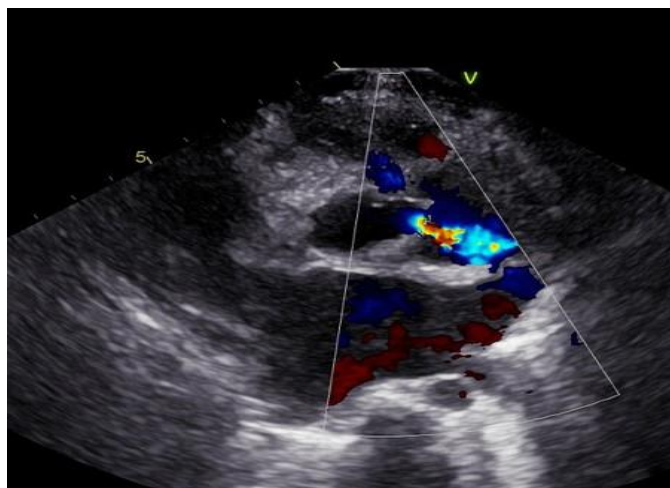
A transthoracic echocardiogram (TTE) disclosed a ruptured sinus of Valsalva aneurysm dissecting into the interventricular septum.

There was an orifice of communication measured at 5mm with flow within the aneurysm.

- The sinus of Valsalva aneurysm appeared to be originating predominantly from the right coronary sinus, although the non- coronary sinus was not entirely excluded.
- There was an obvious disruption of the sinus wall, indicating rupture.
- Color Doppler showed turbulent flow from the aorta into the IVS through an orifice measuring approximately 5mm in diameter.

TOE showing the ruptured aneurism

- There was a probable compression effect seen towards the region of the conduction system which correlated clinically with the patient's complete atrioventricular block.
- No obvious intracardiac shunting was seen on color Doppler, indicating that the rupture was contained within the septum and was not communicating with the right ventricle.



TOE: Orifice of communication between the aneurysm and the dissected septum = 5 mm

AngioTDM:

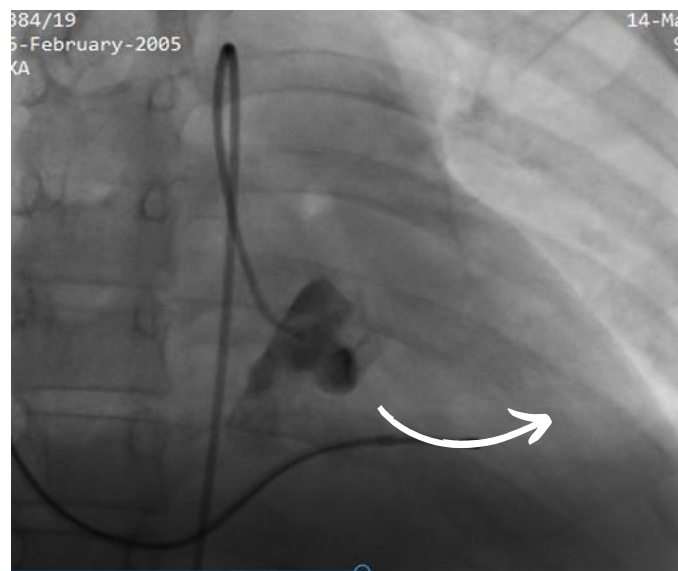
Angiography utilizing computed tomography (AngioCT) provides detailed anatomical and spatial resolution,

- The AngioCT clearly depicted the rupture point of the RSVA with the contrast extending into the IVS, confirming the communication between the aortic root and the IVS.
- The dissection within the septum was evident as a contrast-filled channel, revealing the extent and direction of the dissection. Measurements were consistent with the echocardiographic findings, showing the orifice of communication between the RSVA and the IVS to be approximately 5mm in diameter.
- The AngioCT provided a detailed view of the surrounding cardiac structures. The proximity of the RSVA to the conduction system, especially the AV node and the bundle of His, indicated the likely mechanism for the patient's complete AV block.
- No evidence of compression or involvement of adjacent coronary arteries was seen

Aortic angiography:

Using the Seldinger technique, a catheter was advanced into the ascending aorta. Once in place, a bolus of iodinated contrast agent was rapidly injected, and real-time fluoroscopic images were captured in various angulations.

- An outpouching, consistent with an aneurysm, was visible from the right coronary sinus. As the contrast filled the aortic root, an extravasation was evident, representing the rupture of the sinus of Valsalva.
- There was a direct flow of contrast from the right coronary sinus into the IVS, confirming the communication between the ruptured aneurysm and the IVS.
- The flow dynamics exhibited a high pressure jet stream during systole from the aorta into the septum, which became diffused within the septal dissection.
- The orifice through which the contrast flowed from the RSVA to the IVS was clearly visualized.
- Using calibration tools in the fluoroscopic system, the diameter of the orifice was approximated to be around 5mm, consistent with previous imaging modalities.

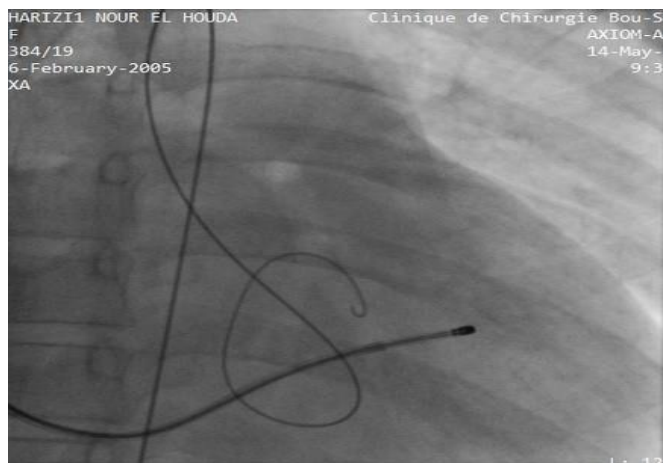


Angiography Detailed anatomical confirmation of the rupture of sinus of Valsalva and showing the contrast going inside the dissected interventricular septum

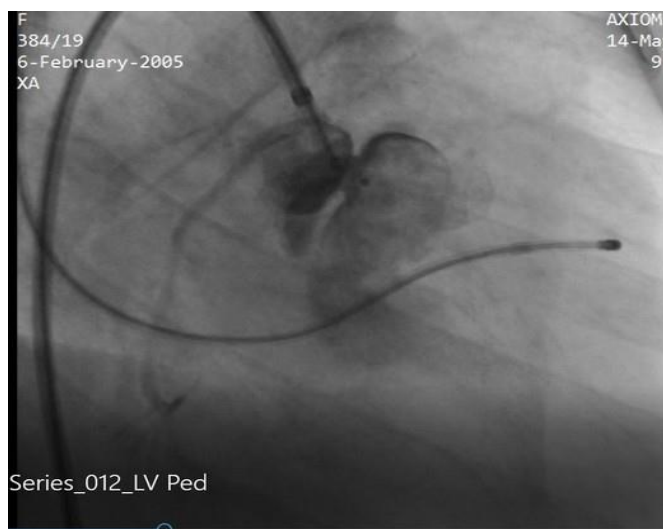
Percutaneous Occlusion:

Pre-procedural Planning:

1. **Imaging:** Prior detailed imaging with echocardiography, AngioCT, and fluoroscopic angiography is essential to understand the anatomy, size, and exact location of the rupture.
2. **Choice of Device:** Based on imaging, an appropriate occluder device, such as a VSD muscular occluder, is selected. The size is typically chosen to be slightly larger than the measured defect to ensure a snug fit.



the first disk opened inside the dissected septum



Angiography prior to the device release showing a good position

- First, the left ventricular disc (or part of the device meant to sit on the left side) is deployed. The sheath is then pulled back, deploying the right aortic disc (or part meant to sit on the aorta side), thereby sandwiching the defect between the two parts of the device.
- Correct positioning is ensured with fluoroscopy and echocardiography. No residual flow should be evident across the defect.

1. Device Release:

Once the operator is satisfied with the device's position and function, it is released from the delivery system. The delivery sheath is then carefully removed.

2. Post-deployment Imaging:

A repeat angiography is performed to ensure no residual shunt, proper seating of the device, and to assess nearby structures for any interference or impingement. Echocardiography may be used to assess the device position and cardiac function.

3. Closure:

- Hemostasis at the femoral access site is achieved using manual compression, vascular closure devices, or suture techniques.
- The patient is then transferred to a recovery area for

monitoring.



Final angiography with complete occlusion of the communicating orifice

- The patient is transferred to a recovery room for monitoring.
- Vital signs, vascular access site, and overall patient well-being are monitored closely.
- After a few hours and ensuring no complications, the patient can be shifted to a regular room.

Follow-up:

Echocardiography is performed within 24 hours to confirm the device's position and assess the false aneurysm's status. Periodic follow-up with imaging, usually echocardiography or CT angiography, is scheduled to monitor the long-term position of the device and the status of the aneurysm.

Discussion:

The decision to treat and the mode of intervention hinge upon the severity of the rupture, the presence of complications such as arrhythmias or heart failure, and the risk profile of the patient.

Therapeutic Choices:

Conservative Management: Some asymptomatic SVA without rupture might be managed conservatively with regular monitoring. However, once rupture occurs, watchful waiting increases the risk of complications, making intervention generally necessary

Surgical Repair:

Open Heart Surgery: Traditionally, ruptured SVA was repaired through open-heart surgery involving cardiopulmonary bypass. The ruptured sinus is usually closed with a patch, and any resultant aortic regurgitation can also be addressed.

Advantages: Definitive treatment, especially for larger ruptures or those with complicating factors such as significant aortic regurgitation.

Disadvantages: Invasive nature, longer recovery period, risk of complications including infection, bleeding, and damage to surrounding structures.

Percutaneous Intervention:

Over recent years, percutaneous closure of RSVAs has emerged as a viable option, especially for select cases.

Advantages: Minimally invasive, shorter hospital stay, and quicker recovery. Especially relevant for patients who may not tolerate open-heart surgery well.

Disadvantages: Limited by device sizes and shapes, potential for device malposition or embolization, risk of residual shunt.

Decision-making in this Case:

Given our 12-year-old patient's profile, several factors influenced our decision towards a percutaneous approach:

Age and Size: Pediatric patients, especially those as young as 12, often benefit from minimally invasive procedures due to faster recovery times and less postoperative discomfort.

Rupture Characteristics: The orifice size of 5mm and its location suggested a feasible percutaneous closure.

Presence of AV Block: The AV block likely resulted from compression by the dissecting aneurysm. A successful percutaneous intervention could potentially relieve this compression, offering a chance for conduction recovery.

Technical Expertise: Our interventional team's experience with similar percutaneous procedures, like ASD and VSD closures, meant that we had the requisite skill set for this intervention.

Risks of Open Surgery: Given the location of the rupture and its proximity to the conduction system, open surgery carried risks of further damage to this already compromised region.

Patient and Family Preference: After explaining both procedures, the patient's family preferred the less invasive option.

Conclusion:

Therapeutic choices for a ruptured sinus of Valsalva aneurysm are multifaceted. While open-heart surgery

remains a standard treatment in many cases, percutaneous closure offers a less invasive option for suitable candidates. In our patient's case, given her age, the characteristics of the rupture, the potential reversibility of her AV block, and the expertise of our team, the percutaneous approach emerged as a favorable choice. The decision should always be individualized, considering the patient's unique clinical scenario and available expertise. This case underscores the importance of considering a structural cardiac anomaly in young patients presenting with conduction disturbances. It also highlights the efficacy and safety of percutaneous interventions in managing such conditions.

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