

#ASEchoJC Twitter Chat

Tuesday, November 15, 2022 – 8 PM ET

- [Mysterious Infantile Cyanosis: An Imaging Case Series](#) (CASE, October 2021)
- [Fatal Sinus of Valsalva Aneurysm and Dissection into the Left Ventricle With Extension to the Interventricular Septum: A Challenging Diagnosis](#) (CASE, December 2021)

Authors

- Nilda Espinola Zavaleta, MD (@nilda_zavaleta)
- Sudeep Sunthakar, MD (@SunthakarMD)

Moderators:

- Kelly Boegel, ACS, RCCS, RCS, FASE (@boegel_kelly)
- Enrique Garcia-Sayan, MD, FACC, FASE (@EGarciaSayan)
- Ritu Thamman, MD, FASE, FACC (@iamritu)
- Vincent L. Sorrell, MD, FASE (@VLSorrellImages)

Enrique Garcia-Sayan, MD, FACC, FASE Tweetorial: 11-14-2022

<https://twitter.com/EGarciaSayan/status/1592125119589416963>

Kelly Boegel, ACS, RCCS, RCS, FASE, Clinical Lead Cardiac Sonographer

Tweetorial: 11-14-2022

https://twitter.com/boegel_kelly/status/1592073738098724871

Introduction and Welcome: 🌟 Welcome to tonight's

#ASEchoJC! 🌟 2 @CASEfromASE discussions w/
authors @SunthakarMD @nilda_zavaleta, EiC
@VLSorrellImages & co-moderators @iamritu
@boegel_kelly

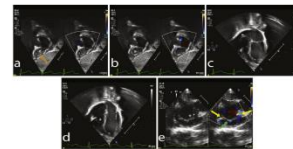
<https://bit.ly/3UvHuPX> ? 1-5

<https://bit.ly/3E3esQP> ? 6-10 (English+Spanish)

👉 Use #ASEchoJC in all Tweets

Case 1 Presentation

A 6-week-old with cyanosis presented as a transfer from an outside hospital with serial right-sided chest tubes with a plan for *Diaphragm-Thymus* transfer to augment pulmonary blood flow. Preoperatively, he was followed by a fetal cardiologist for a diagnosis of hypoplastic right ventricle (RV). Following delivery, he was observed in the neonatal intensive care unit (NICU), where he was able to maintain oxygen saturations in the 80s. He was discharged after 2 days of observation. Over the course of the following 5 weeks, his oxygen saturations began to drift lower into the 70s. At 5 weeks old, he began to have difficulty with feeds and was found to have episodes of oxygen saturations in the 50s. The patient was transferred to our institution for surgical management. Upon arrival, an echocardiogram demonstrated a normal sized right atrium and an undifferentiated and mildly hypoplastic RV. The tricuspid valve annulus measured normal, but the pulmonary valve annulus (Z score, -2.4) and branch pulmonary arteries (left pulmonary artery Z score, -2.9, and right pulmonary artery Z score, -3.0) were mildly hypoplastic. A patent foramen ovale (PFO) was present with predominantly right-to-left shunting explaining the cyanosis. A hypoplastic structure was seen in the right atrium that at times overlapped the tricuspid valve. Color Doppler demonstrated limited inflow across the tricuspid valve due to the presence of this membrane (Figure 1A-4). Videos 1 and 2. An agitated saline contrast injection study confirmed the right-to-left atrial-level shunt (Video 3). The following day, the patient was taken to the operating room for membrane resection. A membrane with attachments to the superior vena cava (SVC), Eustachian valve, and tricuspid valve annulus was resected, consistent with a diagnosis of CTD. A single hole was noted in the specimen (Figure 2). Corroborating images obtained with intraoperative transesophageal echocardiography (TEE) (Figure 3, Video 4). Following membrane resection, while still in the operating room, the chest's oxygen saturations were 95%. In addition, there was improvement in filling of the RV. He recovered in the hospital and was discharged home on postoperative day 4.



Case Presentation

A 42-year-old farmer with no relevant medical history came to the emergency department with dyspnea, chest pain, and altered mental status. His symptoms had limited his daily activities for the past 6 months until they became incapacitating. At admission he had tachycardia (heart rate 116 beats/min), tachypnea (respiratory rate 28 breaths/min), hypotension (blood pressure 104/60 mm Hg), and desaturation (oxygen saturation 87%), as well as increased pale capillary refill > 2 sec, distal cyanosis, and generalized edema. A grade II/IV diastolic murmur on the right parasternal line and a Glasgow Coma Scale score of 7 were observed.

Echocardiography showed sinus rhythm and a left bundle branch block (Figure 1A). Troponin I level was within the normal range, excluding a myocardial infarction. Chest radiography showed grade II cardiomegaly and a right basal pleural effusion (Figure 1B). Transthoracic echocardiography revealed left ventricular global hypokinesia and dilation of the four chambers (Figure 2) with left ventricular end-diastolic volume of 105 mL/m² and end-systolic volume of 61 mL/m². There was systolic dysfunction, with a left ventricular ejection fraction of 42% determined using the modified Simpson method and classic cyphoid with a restrictive pattern (E/A ratio 1.8 and deceleration time 100 msec; Figure 3). There was also moderate mitral regurgitation (vena contracta 0.55 cm) and severe tricuspid regurgitation (vena contracta 0.71 cm), and elevated systolic pulmonary arterial pressure (75 mm Hg; Figure 4). A dissection of the right coronary sinus of Valsalva into the left ventricle (Figure 5, Video 1-3) and extending into the left outflow tract and the interventricular septum with severe aortic regurgitation (pressure half time 170 msec; reverse holodiastolic flow pattern in the abdominal aorta) was also found (Figure 6). No ventricular septal defect (VSD) was detected.

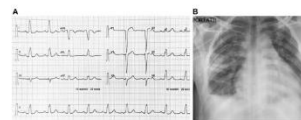


Figure 1 Initial evaluation. A 42-year-old man with progressive dyspnea and exertional chest pain for 6 months. (A) Twelve lead electrocardiography with evidence of a left bundle branch block. (B) Posteroanterior chest radiography that shows grade II cardiomegaly with a cardiothoracic index of 0.63, right basal pleural effusion on the right hemithorax, and indirect signs of pulmonary capillary venous hypertension.

ASE'S TWITTER JOURNAL CLUB

ASE AMERICAN SOCIETY OF
ECHOCARDIOGRAPHY
Sound Saves Lives

Tuesday, Nov. 15, 2022
8:00 to 9:00 PM (ET)

JOIN THE DISCUSSION ON:

2022 ShowCASE Finalists

- » Mysterious Infantile Cyanosis: An Imaging Case Series
- » Fatal Sinus of Valsalva Aneurysm and Dissection into the Left Ventricle With Extension to the Interventricular Septum: A Challenging Diagnosis

Guest Authors Nilda Espinola Zavaleta, MD (@nilda_zavaleta) and Sudeep Sunthakar, MD (@SunthakarMD) and CASE Editor-in-Chief Vincent L. Sorrell, MD, FASE (@VLSorrellImages) will join ASE Twitter Journal Club Moderators Kelly Boegel, ACS, RCCS, RCS, FASE (@boegel_kelly); Enrique Garcia-Sayan, MD, FACC, FASE (@EGarciaSayan); Ritu Thamman, MD, FASE, FACC (@iamritu)

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The ASE Twitter Journal Club will answer your questions!
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Q1:

Question 1 #ASEchoJC

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Since 1963

What is the embryologic origin of cor triatriatum dexter?

11/15/22 @SunthenkarMD @nilda_zavaleta @VL.Sorrellimages @boegel_kelly @EGarciaSayan @iamritu

A1 Notable Responses:

@EGarciaSayan: ▲ CTD due to complete persistence of the R valve of the sinus venosus, normally reabsorbed during embryogenesis.

▲ More common (partial) variants of this phenomenon include prominent Eustachian valve & Chiari network.

👉 See this JASE review: <https://bit.ly/3hBJV5b>

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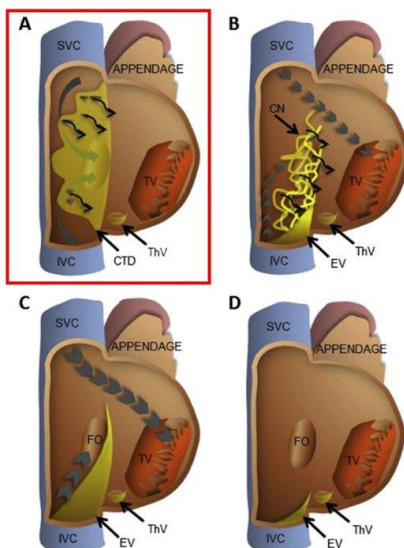


Image from: J Am Soc Echocardiogr. 2016 Mar;29(3):183-94.

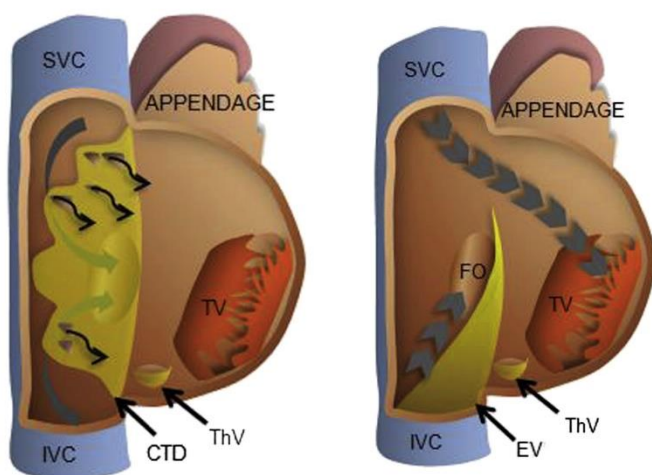
@AntonioBarros_: What is the embryologic origin of cor triatriatum dexter?

A: right sinus venosus

@AntonioBarros_: In CTD, a persistent embryonic right sinus venosus valve becomes a septant membrane that separates the VC & coronary sinus from the right atrial appendage and tricuspid valve. Septation can be partial or complete.

@boegel_kelly: Embryologic origin of CTD is the maldevelopment of the right sinus of venosus which forms the Thebesian and Eustachian valves

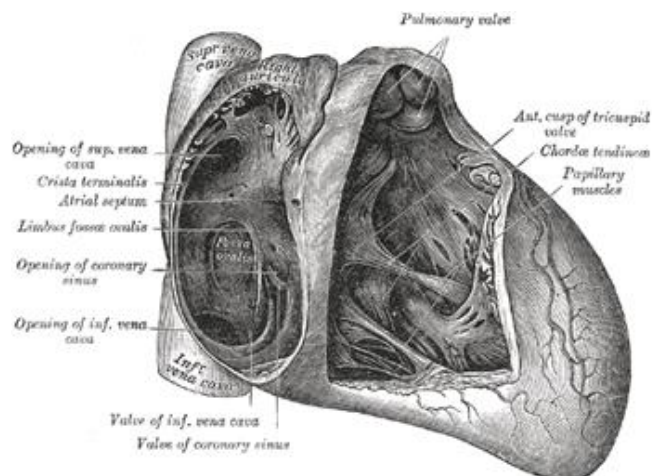
@iamritu: Abnormal persistence of right valve of sinus venosus results in cor triatriatum dexter a prominent EV can mimic CTD if RA appears septated. But in CTD, partitioning membrane is attached to interatrial septum vs prominent EV no such attachment <http://bit.ly/3TFMZdZ>



@SunthankarMD: CTD = maldevelopment of the right valve of the sinus venosus, which forms the Thebesian and Eustachian valves. Thus, the membrane typically attaches to the SVC, Eustachian valve, and the inferior/posterior atrial septum near the coronary sinus.

@VLSorrellImages: FYI, Bartolomeo Eustachio (c 1550-1574) was an Italian anatomist who, like many of his generational peers, was unable to publish much of his work which derived from carefully performed human dissections - a controversial practice in the eyes of the church.

@VLSorrellImages: Henry Gray 1918:



@EGarciaSayan:

▲ Incomplete variants of CTD have been described as "forme frustre" in

@CASEfromASE report by @ray_stainback et al

▲ ff-CTD membrane attached to atrial septum forms a Y sign but does not cause obstruction

▲ Can impact transcatheter procedures

👉 <https://bit.ly/3WYs74s>

Question 2:

Question 2 #ASEchoJC

What is the clinical presentation of cor triatriatum dexter? Is it different in children and adults?

11/15/22 @SunthankarMD @nilda_zavaleta @VL.Sorrellimages @boegel_kelly @EGarciaSayan @iamritu

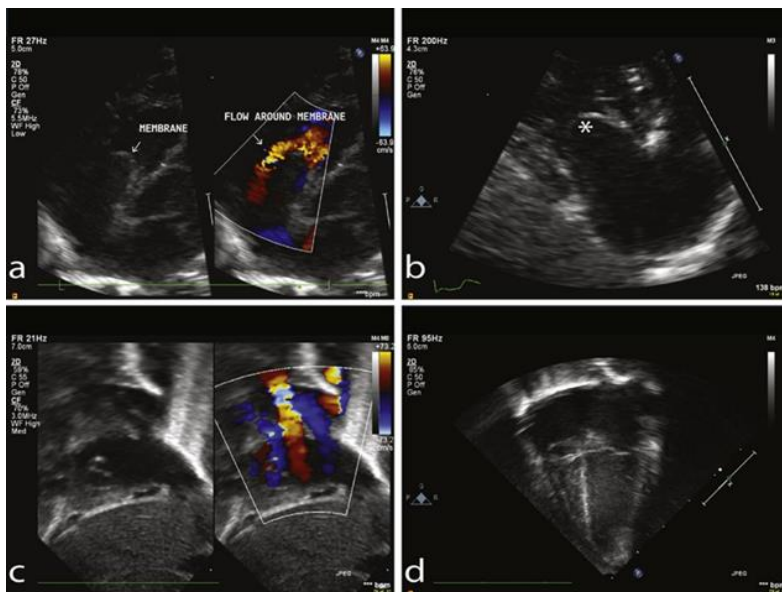
A2 Notable Responses:

@EGarciaSayan:

▲ CTD has variable clinical presentation depending on degree of RA partitioning, fenestrations & associated lesions, ASD, PFO

▲ Can be asymptomatic or cause R HF & ↑ CVP

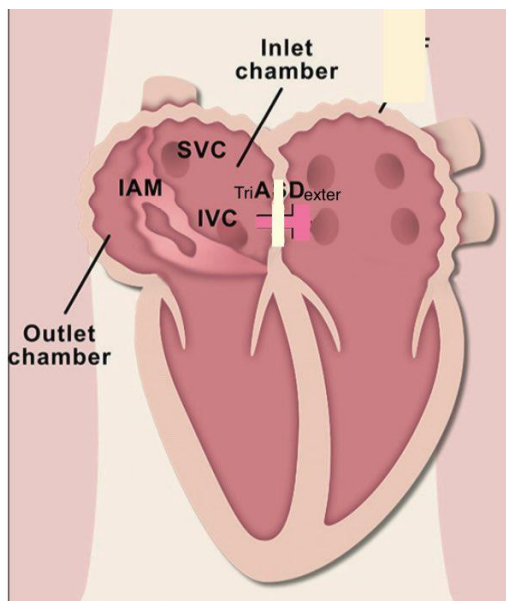
▲ May result in cyanosis and severe hypoxemia such as in this case by @SunthankarMD



@iamritu: varies depending on degree of RA partitioning/ septation

If septation mild, the condition asymptomatic/ incidental finding in vast majority

If severe septation Can get right-sided HF elevated CVP due to obstruction of tricuspid valve, RVOT or IVC



@SIwa23288585: Our case. A full-term neonate presented with cyanosis only when sleeping, which was considered due to a prominent eustachian valve, directing blood flow from the IVC to the LA through the PFO resulting in interatrial R-L shunting.

<https://pubmed.ncbi.nlm.nih.gov/19488952/>

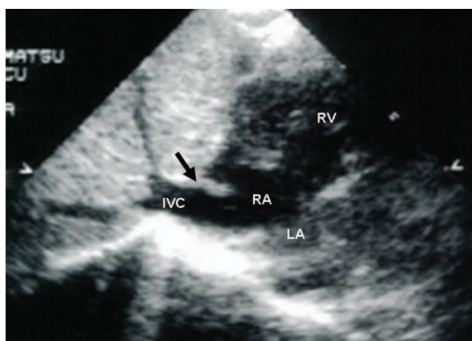
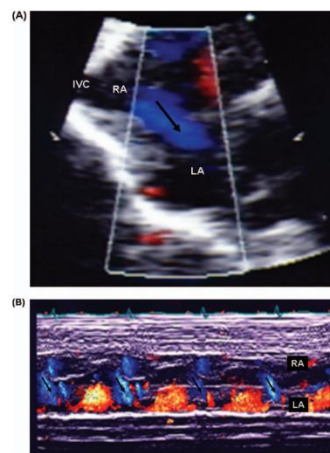


Figure 1. Two-dimensional echocardiographic image from subxiphoid view. A prominent eustachian valve was observed in the right atrium (arrow). IVC, inferior vena cava; RA, right atrium; RV, right ventricle; LA, left atrium.



Doppler echocardiograms from subxiphoid view. (A) Colour Doppler echocardiography revealed interatrial shunt flow from the IVC to the LA, indicated by a black arrow. (B) 1-mode indicated the shunt flow induced in diastole, which was in atrial systole. Arrows indicate the direction of flow.

@SunthankarMD: Presentation of CTD is variable for pediatric pts.

Pts may go undiagnosed in childhood or present w/ significant hypoxia (assuming ASD present & R→L) as a newborn

🔑 feature determining the timing and severity of presentation is the degree of systemic venous

@SunthankarMD: Since lesion is a congenital malformation, adults have lived their whole lives with membrane present & thus must have been asymptomatic if not diagnosed in infancy.

Our experience → degree of systemic venous obstruction lessens over time rather than increase.

@SunthankarMD: Case reports of CTD dx in adults describe it incidentally found during 1) echo or cross-sectional imaging for an unrelated reason 2) during a right heart cath when the membrane entangles the catheter or 3) post-mortem autopsy (not cause of death).

@VLSorrellImages: "Degree of septation correlates with intracardiac pressures, RHF"

mild = asymptomatic- incidental finding

severe = high CVP, RH failure, or obstruction to the IVC or RVOT

@VLSorrellImages: Depends on presence of perforations in the membrane, size of perforations, right-to-left shunt & associated malformations.

@AntonioBarros_: The major physiologic consequence is elevation of pressure in the accessory chamber that is proximal to the obstruction.

Question 3:

Question 3 #ASEchoJC

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BRIDGE YOUR LIVES

What associated lesions do we need to look for in patients with cor triatriatum dexter?

11/15/22 @SunthakarMD @nilda_zavaleta @VL_SorrellImages @boegel_kelly @EGarciaSayan @iamritu

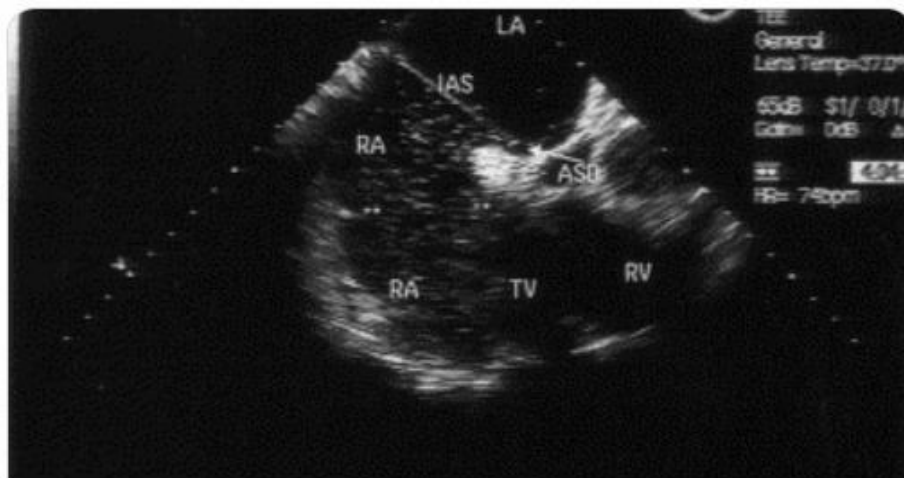
A3 Notable Responses:

@EGarciaSayan:

- ▲ CTD often associated with other R-sided malformations which must be excluded by #EchoFirst
- ▲ ASD, PFO
- ▲ Ebstein anomaly
- ▲ Tricuspid or pulmonic stenosis or atresia
- ▲ Hypoplastic RV

@iamritu: 50% cases assoc w other cardiac anomalies of such as pulmonary stenosis, tetralogy of Fallot, Ebstein anomaly <http://bit.ly/3TETfmf>

D/D prominent Eustachian valve, redundant Chiari's, especially when membrane is thin, mobile, & engaging into tricuspid valve orifice



onlinejase.com

Cor triatriatum dexter, atrial septal defect, and Ebstein's anomaly in a...

Cor triatriatum dexter is a very rare congenital abnormality in which the right atrium is divided into two chambers by a membrane. In this ...

@boegel_kelly: Check for atrial septal defects with right to left shunting as seen in this CTD case in TEE imaging 📌 <https://twitter.com/i/status/1592687772456083456>



@iamritu: When ASD is present with CTD & there's increased atrial pressure (ie from AFib) due to inadequate atrial emptying may **↑** right to left shunting across ASD

@DavidWienerMD: Reverse Lutembacher's physiology

@SunthankarMD: As with all forms of congenital heart disease, a thorough complete study should be performed to assess all chambers, valves, and great arteries and veins.

@SunthankarMD: The 🔑 features to focus on with CTD are size of right sided structures (i.e. tricuspid valve, pulmonary valve, and right ventricle) as well as size of the atrial septal communication. In congenital cardiology we use the mantra "no flow = no grow"

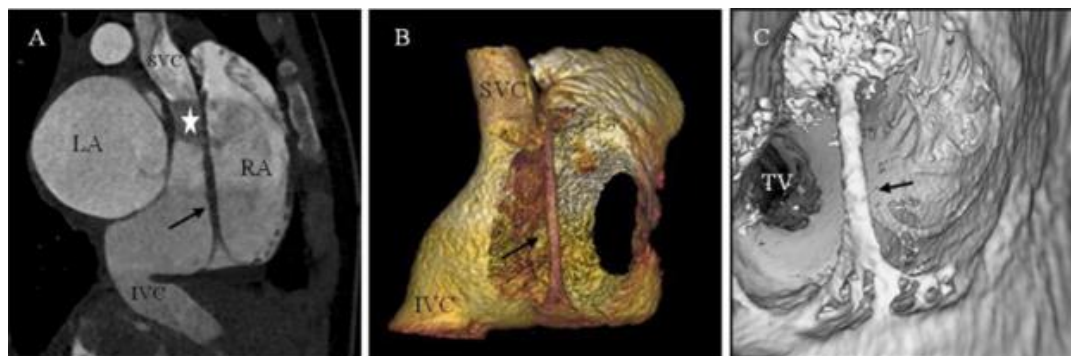
@SunthankarMD: We describe CTD with pulmonary atresia/intact ventricular septum (Case 4 in article). In this scenario, it could be postulated that the obstructive membrane --> tricuspid stenosis, hypoplastic right ventricle, and pulmonary atresia.

@rajdoc2005: No Flow = No Grow!! ❤️ that mantra!

@VLSorrellImages: Here is a rare CASE associated with HCM in a young lady:

https://ars.els-cdn.com/content/image/1-s2.0-S193004332200262X-gr2_lrg.jpg

<https://doi.org/10.1016/j.radcr.2022.03.087>



@VLSorrellImages: In my simple mind:

Since CTD represents an embryologic failure of the right heart, associations to other RH pathology may occur (e.g. Ebsteins) ASD / PFO not uncommon & may lead to hypoxemia or paradoxical emboli

Question 4:

Question 4 #ASEchoJC

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How do we assess the adequacy of fenestrations by #EchoFirst in cor triatriatum dexter?

11/19/22 @SunthakarMD @nilida_zavalata @VLSorrellImages @boegel_kelly @EGarciaSayan @iamritu

A4 Notable responses

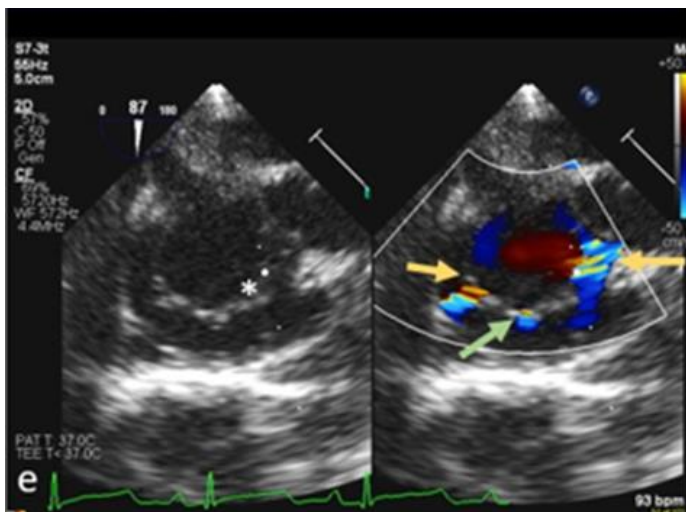
@boegel_kelly: Take time to carefully evaluate the adequacy of fenestrations:

- ✳ use all available windows
- ✳ use optimized color Doppler
- ◆ Reduce color gain
- ◆ color scale set 50-70Hz
- ✳ look for turbulence in color Doppler
- ✳ spectral Doppler gradients
- ✳ 3D imaging
- ✳ TEE

@iamritu: based on number & size of fenestrations in fibro-muscular membrane
@SunthakarMD

- 💡 absence of connection between the two chambers,
- 💡 one or few small openings in the intra-atrial membrane
- 💡 Or accessory chamber communicates widely by a large single opening

@SunthakarMD: There are three main ways to assess adequacy of fenestrations by echo: 1) Color flow Mapping to identify number and size of fenestrations (green arrow) within the membrane. Also, it is crucial to look for flow that circumvents the membrane (yellow arrow).



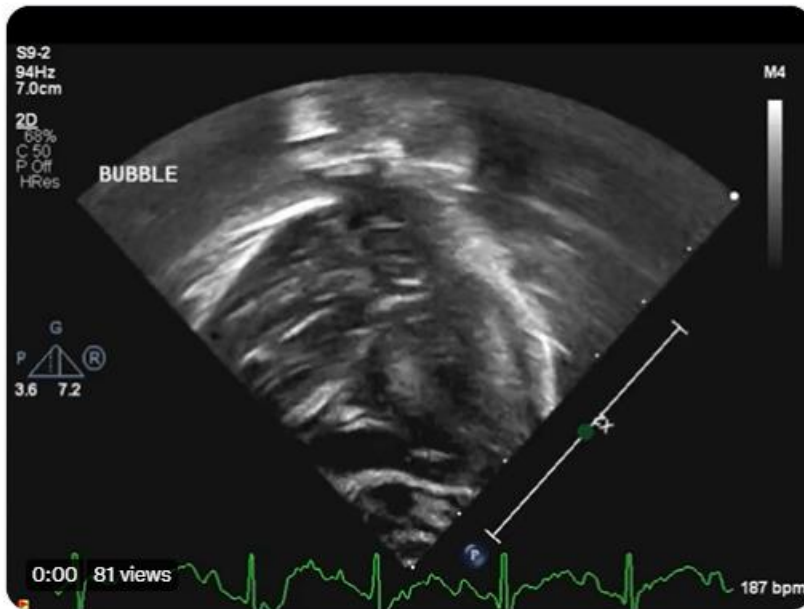
@SunthakarMD: Pulse wave Doppler to detect prograde flow across the tricuspid valve

@SunthakarMD: Look for secondary features of systemic venous obstruction which may include
--> TV, RV, PV hypoplasia

--> significant right to left atrial level shunting.

@SunthankarMD: And a bonus 4th!

4) Perform an agitated saline study to look for relative flow across the tricuspid valve versus the atrial septal communication. Here you see significant R-->L atrial shunt in the setting of significant CTD obstruction <https://twitter.com/i/status/1592690040706314241>



@iamritu: How often do you use #UEA to help characterize fenestrations in CTD?

@boegel_kelly: I'd love to see some images of #UEA to help characterize fenestrations.

Question 5:

Question 5 #ASEchoJC

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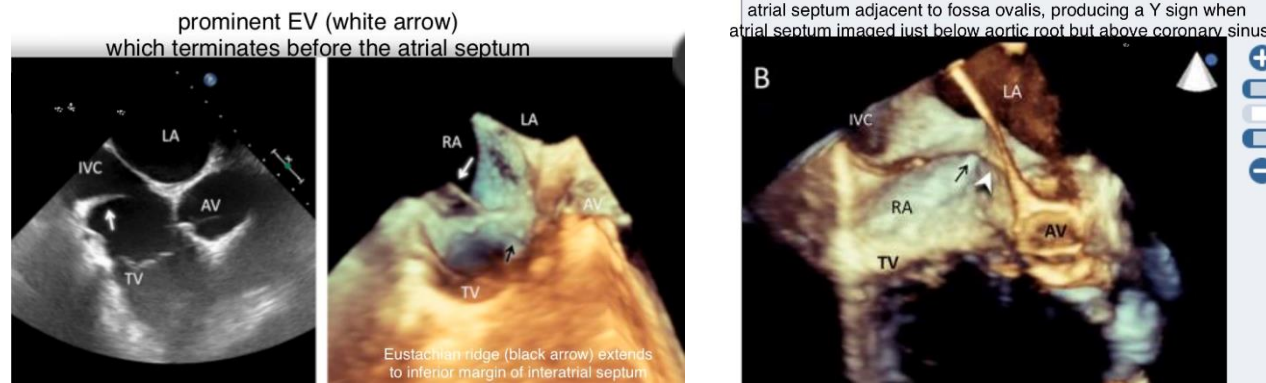
When is a surgical intervention for cor triatriatum dexter needed?

11/15/22 @SunthakarMD @niida_zavaleta @VL_SorrellImages @boegel_kelly @EGarciaSayan @iamritu

A5 Notable responses

@iamritu:

<http://bit.ly/3g507LY> remember to look out for CTD before doing any interventions CTD remnant is usually close to an ASD, if present, & may make percutaneous closure of ASDs, tricky. it can be confused w prominent Eustachian Valve(not attached to interatrial septum)



@SunthakarMD: As with most things in medicine, particularly more rare conditions, it really is an individualized approach to determine which patients require surgical intervention with membrane resection.

@SunthakarMD: Over the first 2–3 months of life, right-to-left atrial-level shunt in CTD may decrease.

This is likely due to a combination of somatic growth reducing the effective obstruction of the membrane over time and decreasing pulmonary artery pressures.

@SunthakarMD: Observing pts through this transitional period is encouraged if patient is not having persistent O₂sat<80% or episodic significant desaturations.

Those who may be good candidates for observation have adequate fenestration size and prograde flow across the TV

@SunthakarMD Nevertheless, clinical findings are 🏰. Treatment should be based on the patient's clinical appearance, with saturations as the primary outcome measure.

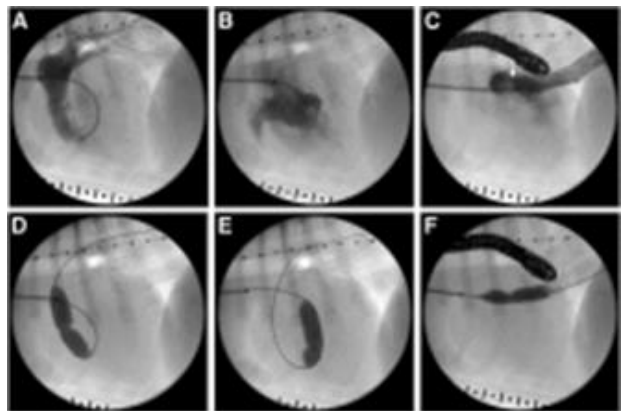
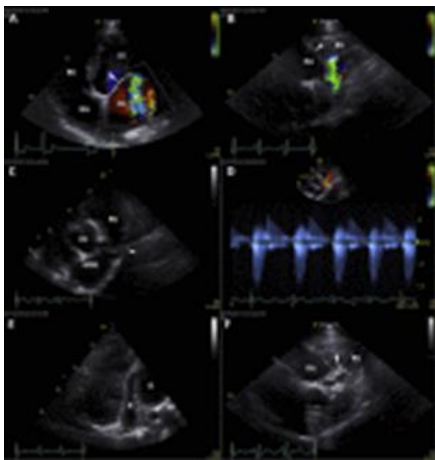
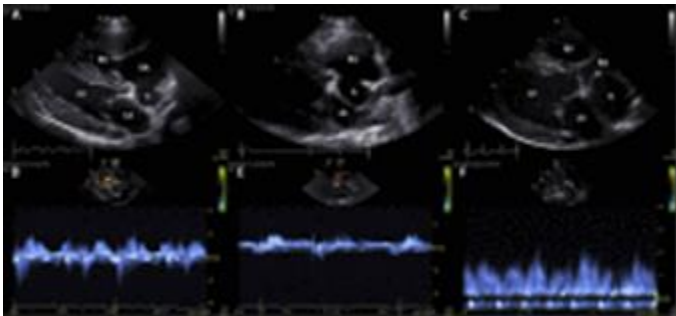
@SunthakarMD: Resection when:

- persistent hypoxia that does not improve with age
- hemodynamically significant right-to-left shunt causing hypoplasia of the right-sided structures and/or cyanotic spells

@SunthakarMD: In one patient, oxygen saturations were 78% during the pre-operative TEE and 99% during the post-operative TEE after this 🏰 was resected.



@VLSorrellImages: @CASEfromASE has example of percutaneous repair:



Question 6:

Question 6 #ASEchoJC

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2024 Fall Meeting

**How often do SOVA rupture,
and into which chambers?
What are other complications
associated with SOVA?**

11/15/22 @SunthanKarMD @nilda_zavaleta @VL.SorrellImages @boegel_kelly @EGarciaSayan @iamritu

A6 Notable responses

@nilda_zavaleta: An analysis of a published case series by Chu et al. found that the incidence of SOVA rupture is approximately five times higher in East Asian patients than in westerners. The right coronary sinus is most frequently affected, followed by the non-coronary sinus

@nilda_zavaleta: Rupture of the aneurysm most often occurs into the RV, followed by the right atrium and rarely into the left ventricle, pulmonary artery, or interventricular septum

@nilda_zavaleta: Rupture of the right and non-coronary sinuses typically results in communication between the aorta and the RVOT or the aorta and the RA. Left SOVA rupture is clinically less significant, with resultant communication between the LA and LVOT

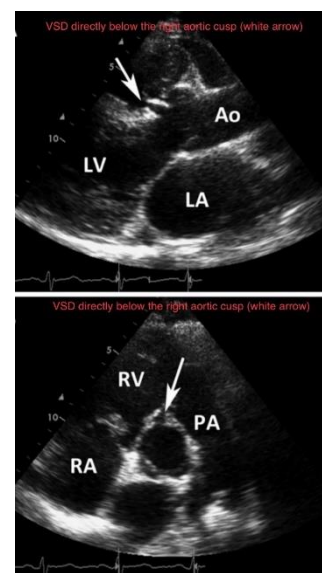
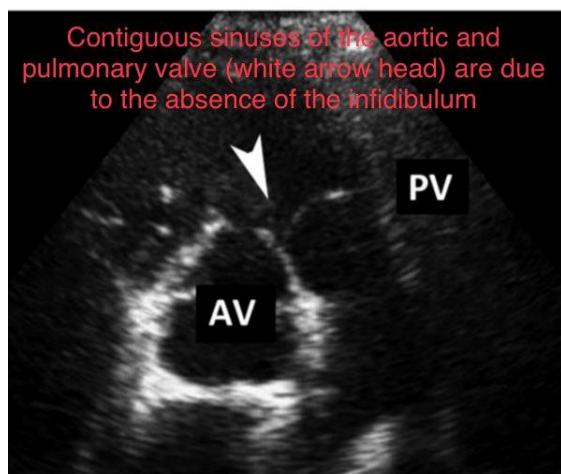
@nilda_zavaleta: The case presented is a right SOVA complicated with septal and anterior wall dissection

@rajdoc2005: Agree. Just wondering why the incidence of rupture is much higher on the right sided sinus than the left sinus..??

@iamritu: Maybe because of the anatomy of the sinuses of Valsalva? This congenital SOVA caused by weakness at juncture of aortic media & the annulus fibrosus is it weakest near the right coronary sinus (65%–85%)

@rajdoc2005: Sounds logical. This is very helpful information! 🙏🙏

@iamritu: most common cardiac defect with SVA is VSD supracristal/ infundibular, when an SVA is in close proximity to a VSD, an unruptured SVA may be misdiagnosed as an SVA with rupture, as opposed to an intact SVA with a coexisting VSD lacks muscular conal septal support in RVOT



@nilda_zavaleta: Una serie de casos publicados por Chu et al. encontró que la incidencia de ruptura de SOVA es aproximadamente cinco veces mayor en pacientes de Asia del Este que en Occidente. El seno coronario derecho es el más frecuentemente afectado, seguido por el seno no coronario

@nilda_zavaleta: La ruptura del aneurisma ocurre con mayor frecuencia en el VD, seguido de la AD y rara vez en el VI, la AP o el tabique interventricular

@nilda_zavaleta: La ruptura de los senos derecho y no coronario generalmente resulta en la comunicación entre la aorta y el TSVD o la aorta y la AD. La ruptura de ASDV izquierdo es clínicamente menos significativa, con comunicación entre la AI y el TSVI

@nilda_zavaleta: El caso presentado es un aneurisma del seno de Valsalva derecho complicado con disección septal y de la pared anterior

@EGarciaSayan:

▲ En serie de Mayo Clinic: aneurismas del SV derecho 70% (usualmente fistula al VD), no-coronario 25% (usualmente fistula a AD), izquierdo es poco común (5%~)

▲ Edad media 45, 63% masculino

📖 Am J Cardiol 2007; 99: pp. 1159-1164: <https://bit.ly/3UR3wfM>

Sinus of Valsalva Aneurysms—47 Years of a Single Center Experience and Systematic Overview of Published Reports

Sherif Moustafa, MD, MSc^a, Farouk Mookadam, MD, MSc^{b,*}, Leslie Cooper, MD^c, Guleid Adam, MD^c, Kenton Zehr, MD^d, John Stulak, MD^d, and David Holmes, MD^c

A retrospective study was undertaken to review demographic data, clinical presentation, outcomes, and long-term results of surgical repair of sinus of Valsalva aneurysms (SVAs). SVAs are a rare anomaly. Surgery is the treatment of choice. A retrospective review of an institutional database identified 86 patients who underwent SVA repair from 1956 to 2003. Follow-up ranged from 3 months to 40 years. The median age was 45 years (range 5 to 80). Approximately 44% of the patients had associated aortic regurgitation. Ruptures occurred in 34% of patients. The predominant fistula was from the right sinus of Valsalva to the right ventricle. Most (65%) were diagnosed by echocardiography, and the remaining (35%) were diagnosed on cardiac catheterization. All subjects underwent SVA repair. Seventy-two patients (84%) underwent other cardiac procedures at the time of aneurysm repair. Six patients (7%) died perioperatively, and the actuarial 10-year survival rate was 63%. In conclusion, echocardiography is the most frequently used diagnostic tool. The most common site of the aneurysm was the right coronary sinus. The concomitant surgical repair of associated ventricular septal defect, atrial septal defect, and the aortic valve is often required. Elective surgical repair can be performed with low risk. © 2007 Elsevier Inc. All rights reserved. (Am J Cardiol 2007;99:1159–1164)

Table 3

Site of origin and exit site of ruptured sinus of Valsalva aneurysms

Site of Rupture	Right Coronary Sinus	Noncoronary Sinus	Left Coronary Sinus	Total
Ruptured				
Right ventricle	16	1	0	17
Right atrium	5	6	0	11
Right ventricle and interventricular septum	1	0	0	1
Nonruptured	38	15	4	57
Total	60	22	4	86

@EGarciaSayan:

▲ SOVA can rupture into heart chambers (usually R), cause compression (RV infow or outflow, coronaries) or affect conduction system

▲ R or L HF Sx depending on chamber affected

▲ Rarely rupture into LV, LA, PA, SVC or pericardium, but extracardiac rupture is very rare

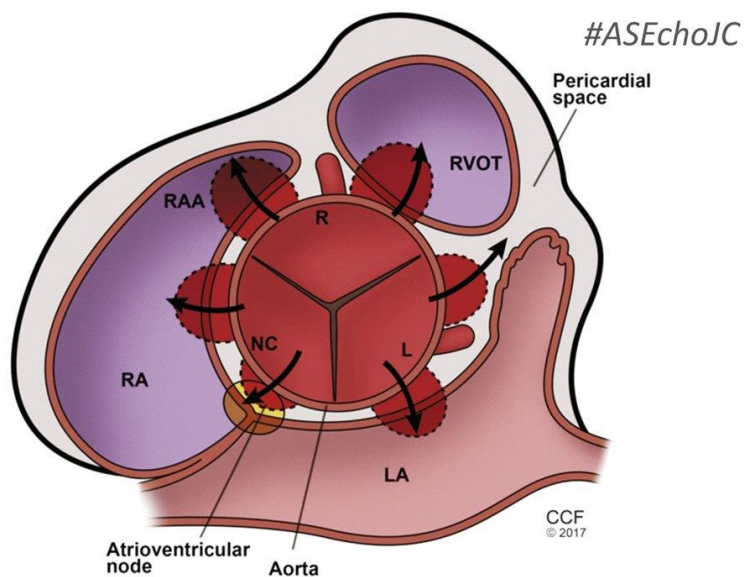



Image from: J Am Soc Echocardiogr. 2020 Mar;33(3):295-312.

Question 7:

Question 7 #ASEchoJC

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Since 1963

What associated lesions do we need to look for in patients with SOVA?

11/15/22 @SurthiankarMD @nilda_zavaleta @VLSorrellImages @boegel_kelly @EGarciaSayan @iamritu

A7 Notable responses

@EGarciaSayan:

▲ Most common associated abnormality are VSD (12-53%), RSOVA or NCSOVA can prolapse through a membranous VSD

▲ Also common association with AR (44% in Mayo Series)

▲ Other associations: PSs, ASD, BAV, tet of Fallot, PDA, coarctation, subaortic membrane.

Table 4
Associated cardiac lesions

Variable	No. (%)
AR	38 (44%)
Ventricular septal defects	27 (31%)
Marfanoid features	9 (11%)
Bicuspid aortic valve	8 (9%)
Mitral regurgitation	8 (9%)
Tricuspid regurgitation	4 (5%)
Atrial septal defect	3 (4%)
Coarctation of aorta	3 (4%)
Ebstein's anomaly	2 (2%)
Tetralogy of Fallot	2 (2%)
Transposition of great vessels	1 (1%)
Patent ductus arteriosus	1 (1%)

Table 5
Associated cardiac procedures

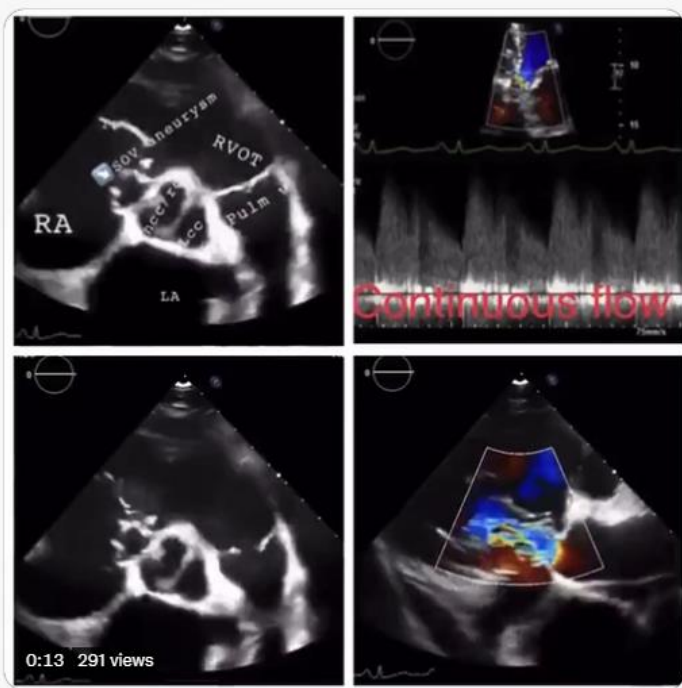
Variable	No.
Ventricular septal defect closure	27
Aortic valve replacement	18
Aortic valve repair	14
Mitral valve replacement	4
Tricuspid valve replacement	3
Tricuspid valve annuloplasty	3
Atrial septal defect closure	3

@nilda_zavaleta: VSD is the most common coexisting cardiac anomaly with SOVA rupture and occurs in 9%–78% of reported patients (30–60%)

@nilda_zavaleta: It is also important to rule out bicuspid aortic valve (15–20%), aortic regurgitation (44–50%), ASD, RVOT obstruction, tricuspid regurgitation, and conduction system involvement

@iamritu: These SOVA are associated w other congenital anomalies, most often VSD (30–60%), bicuspid aortic valve (15–20%), & aortic regurgitation (44–50%)

Here's anterior bicuspid aortic valve fusion RCC & NCC w SOVA rupture <https://bit.ly/325wxKz>



@VLSorrellImages: Here's my \$0.02 regarding BAV & SOVA:

A True BAV (with only 2 SOV) will exert higher stress per sinus than if there are TAV & 3 SOV (increased aneurysm risk).

@VLSorrellImages: Systemic:

Etiology: infection, degenerative diseases, connective tissue disorders, trauma

Autoimmune diseases associated with SOVA

Anatomic:

VSD [$>30\%$] & AR [$\sim 50\%$]

(CoA 4% [be sure to check BP in all extremities] and BAV [15%] rarely)

@EGarciaSayan:

▲ Asociación mas frecuente es defecto septal ventricular (12-53%), aneurismas de SV derecho o no-coronario pueden prolapsar a través de DSV

▲ Insuficiencia aortica ($\sim 44\%$)

▲ Estenosis pulmonar, ASD, BAV, Fallot, PDA, coartación, membrana subaórtica.

@nilda_zavaleta: La CIV es la anomalía cardíaca coexistente más común con la ruptura del ADSV y ocurre en el 9% al 78% de los pacientes reportados (30-60)

@nilda_zavaleta: Además, es importante descartar válvula aórtica bicúspide (15-20%), regurgitación aórtica (44-50%), CIA, obstrucción del TSVD, regurgitación tricúspide y compromiso del sistema de conducción

@EGarciaSayan:

✳ Large single-center (Mayo) series:

▲ RSOVA 70% (most common fistula to RV), NCSOVA 25% (most common fistula to RA), LCSOVA less common (5%~)

▲ Median age 45, 63% male

📄 Am J Cardiol 2007; 99: pp. 1159-1164: <https://bit.ly/3UR3wfM>

Table 3
Site of origin and exit site of ruptured sinus of Valsalva aneurysms

Site of Rupture	Right Coronary Sinus	Noncoronary Sinus	Left Coronary Sinus	Total
Ruptured				
Right ventricle	16	1	0	17
Right atrium	5	6	0	11
Right ventricle and interventricular septum	1	0	0	1
Nonruptured	38	15	4	57
Total	60	22	4	86

@boegel_kelly: 🌟 67%-85% of reported SVAs involve the right coronary sinus with most aneurysms rupturing into the right chambers, rarely in the left

🌟 frequently associated with other CHD lesions

♦ VSDs

♦ bicuspid AoV

♦ aortic regurgitation

@boegel_kelly: 🌟 In patients with existing VSD and a ruptured SVA it can be difficult to distinguish the flows as the SVA shunts overlaps the VSD flow. Take time to properly optimize image and carefully sweep through region to differentiate the two.

Question 8:

Question 8 #ASEchoJC

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Pulse Sound Live

How do we differentiate SOVA from associated VSD or a Gerbode defect?

11/15/22 @SunthakarMD @nilda_zavaleta @VLSorrellImages @boegel_kelly @EGarciaSayan @iamritu

A8 Notable responses

@EGarciaSayan:

- ▲ Discriminating isolated SOVA rupture from SOVA+VSD is challenging
- ▲ Nonruptured SVA near VSD may be misdiagnosed as a ruptured SoVA.
- ▲ SAX view: differentiate thin-walled & fibrous SOVA or prolapsed cusp from thicker myocardium
- ▲ TEE & multi-modality if in doubt

#ASEchoJC

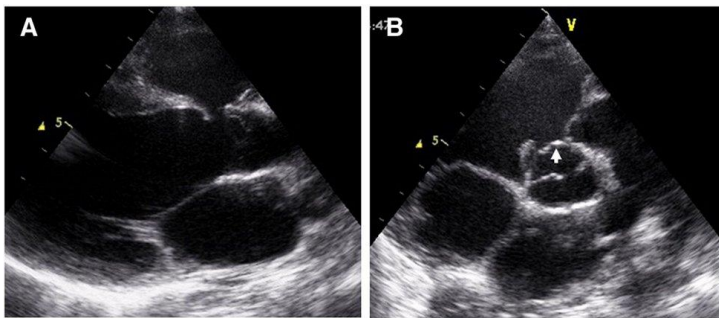


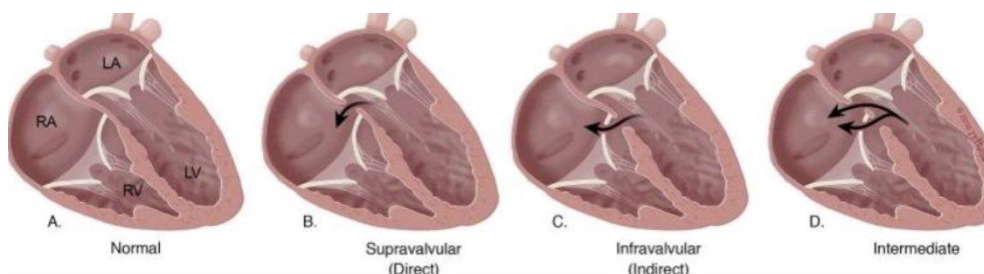
Image from: *Int J Cardiol.* 2014 Apr 15;173(1):33-9.

@nilda_zavaleta: In the presence of a coexisting VSD, the large ruptured SOVA shunt overlaps the VSD flow, which can be difficult to recognize on 2D echo, but 3D echo identifies the two abnormal flows

@nilda_zavaleta: Gerbode defect is a communication from the LV to RA and has shunt from left to right, and it can be divided into congenital or acquired

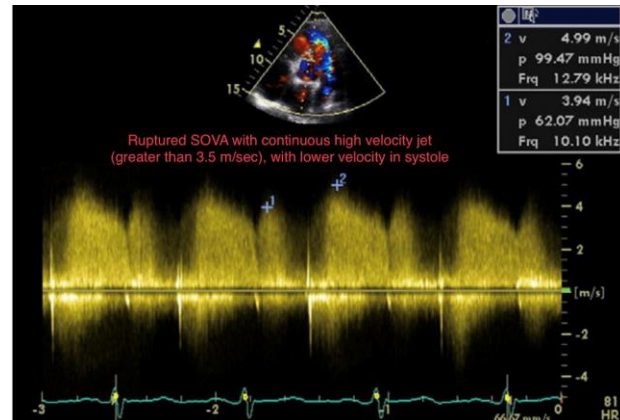
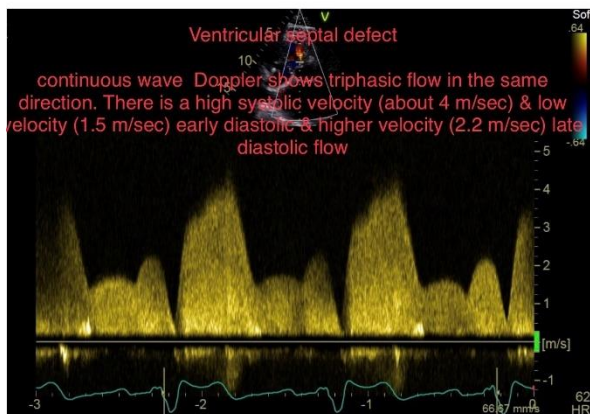
@iamritu: Gerbode Left ventricular to right atrial communications are rare types of ventricular septal defect & present as direct or an indirect types.

LV-RA shunt in systole as left ventricular systolic pressure is greater than right atrial pressure



@iamritu: In an isolated uncomplicated VSD, Doppler shows a high-velocity systolic & a low-velocity diastolic flow

ruptured SOVA will have a left to right shunt with a continuous high velocity jet (greater than 3.5 m/sec) with lower velocity in systole



@rajdoc2005: Great point here. Color flow Doppler can be confusing with all the aliasing from the high flow. CW Doppler shows the directionality of the flow and the timing better!

@rao_sruti: So beautifully explained Dr. Ritu! I always had a hard time differentiating the two with aliasing jets that usually occur. Thank you so much for conveying it across so succinctly! 🙏

@CardiacZhao: Why is the systolic velocity in SOVA lower? Is it because during systole, the pressure gradient between the sinus and RVOT is lower than that during diastole?

@CardiacZhao: My guess is that during systole, the pressure within the Val sinus is lower than that during diastole? Because I think the higher velocity during systole in VSD is probably due to higher LV systolic pressure than diastolic one.

@iamritu: During systole, RSOVA shunt tracks thru RV wall which gets compressed & flow improves in diastole, causing diastolic augmentation

@evolutsapien: Very helpful...physiology always makes pathology understanding more meaningful.

@EGarciaSayon:

- ▲ Diferenciar ASV de ASV + DSV es difícil
- ▲ ASV sin ruptura cerca a DSV puede ser confundido con ruptura de ASV
- ▲ En eje corto, diferenciar la pared delgada y fibrosa de ASV del miocardio mas grueso
- ▲ ETE o multi-modalidad si persisten dudas

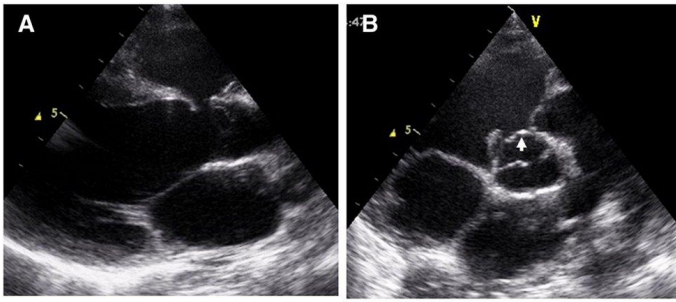


Image from: *Int J Cardiol.* 2014 Apr 15;173(1):33-9.

@nilda_zavaleta: En presencia de una CIV coexistente, el ADSV roto se superpone al flujo de VSD, que puede ser difícil de reconocer en la ecocardiografía 2D, pero la ecocardiografía 3D identifica los dos flujos anormales en la mayoría de los casos

@nilda_zavaleta: El defecto de Gerbode es una comunicación del VI a la AD, tiene cortocircuito de izquierda a derecha y podría clasificarse como congénito o adquirido

Question 9:

Question 9 #ASEchoJC

What are the #EchoFirst or #CVimaging criteria to grade the severity of SOVA for prognosis or surgical indication?

11/15/22 @SunthararMD @nilda_zavaleta @VL.SorellImages @boegel_kelly @EGarciaSayan @iamritu

@EGarciaSayan: imaging in SOVA

▲ Sensitivity >90%

▲ Location, size, relationship / compression of adjacent structures (RV, RVOT, coronaries), abnormal flow into chambers

Evaluate for AR & associated lesions

▲ TEE can help visualize the aorta & ostial coronaries

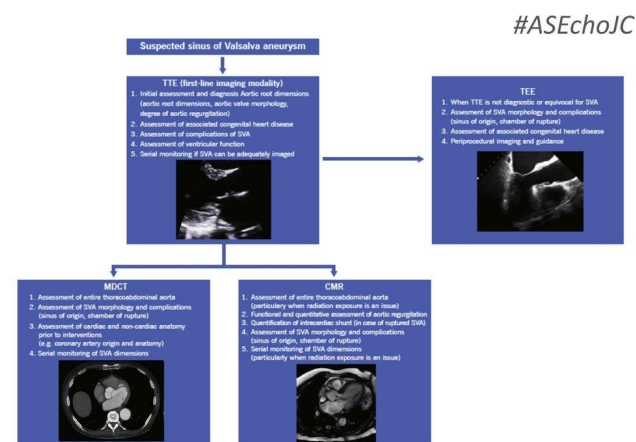


Image from: J Am Soc Echocardiogr. 2020 Mar;33(3):295-312.

@EGarciaSayan: #CVimaging in SOVA

▲ #YesCCT & #WhyCMR can be very helpful

▲ Delineate anatomy & relationship with adjacent structures

▲ Assist with surgical planning

See fantastic SOTA imaging review for SOVA published in @JournalASEcho: <https://bit.ly/3UR8e8D>

@nilda_zavaleta: In the case of a ruptured SOVA, echocardiographic evaluation with color Doppler reveals continuous flow in systole and diastole, flutter of the tricuspid valve as the color jet moves from the aorta to the right chambers of the heart

@nilda_zavaleta: It is important to define the location of the involved sinus and sinotubular portion of the aorta, the coexistence of bicuspid aorta, the involvement of the conduction system

@nilda_zavaleta: the presence and type of shunts, the severity and mechanism of aortic regurgitation, and the vicinity of coronary ostia.

@nilda_zavaleta: Both ruptured and non-ruptured SOVAs can be complicated by aortic regurgitation, occurring in 30-50% of patients. CMR used with multiplanar sequencing has allowed for the evaluation of intracardiac shunts in ruptured SOVAs

@nilda_zavaleta: New cardiac computed tomography modalities allow for accurate diagnosis of even small SOVA's in a dynamic setting. According to the 2010 American Heart Association Guidelines on Thoracic Aortic Disease, surgical repair should be considered in:

@nilda_zavaleta: Those with aneurysms >5.5 cm, >5 cm in those with bicuspid valves, >4.5 cm in the setting of connective tissue disease, or a growth rate of >0.5 cm/year

@nilda_zavaleta: Surgical intervention is recommended for a ruptured SOVA and/or a SOVA with associated intracardiac abnormalities such as VSD or significant aortic regurgitation. Unruptured but symptomatic or enlarging SOVA should also be considered for surgical repair

@iamritu: what's SVA Diameter?

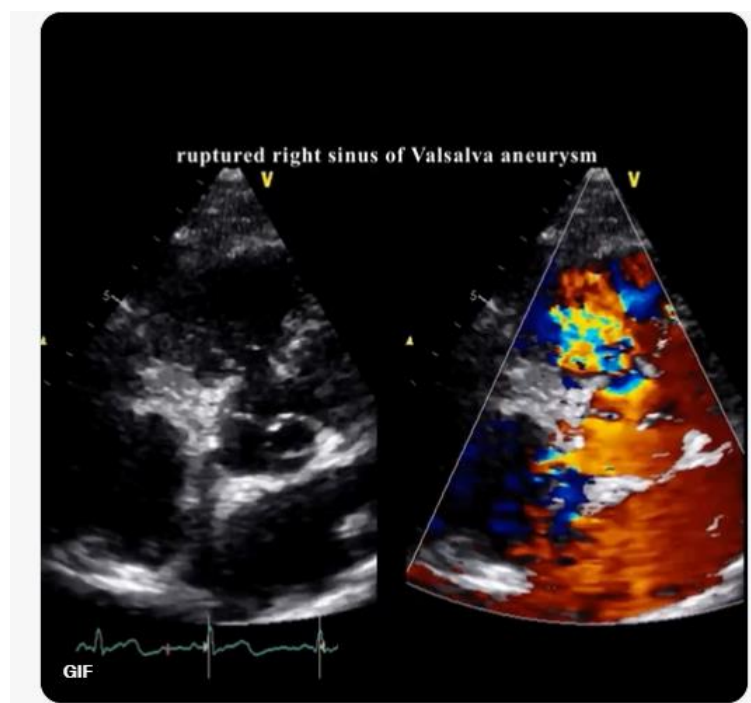
measure SVA diameter perpendicular to long axis of ascending aorta PLAX view at end diastole

What is the origin of SVA? (NC/R/L CC)

How many cusps are involved?

Are any adjacent cardiac chambers compressed by SVA?

<https://twitter.com/i/status/1592697616940535808>



@Ahmed43101178: ● SV Aneurysm #echofirst diagnosis and management

Nice summary by @EGarciaSayan

@EGarciaSayan: ▲ Diámetro de SV en Eco se obtiene en el eje largo paraesternal perpendicular al eje aórtico de borde principal a borde principal (≠ CT/CMR)

▲ Considerar edad, superficie corporal, género, y estandarizar

▲ 3 métodos en la nueva #EchoGuideApp <https://asecho.org/echoguide/>

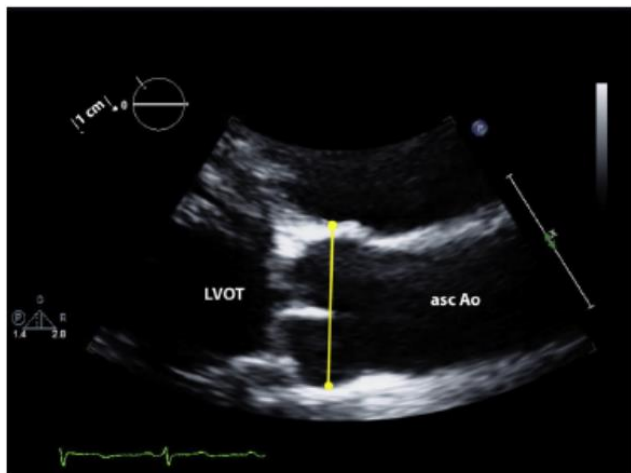
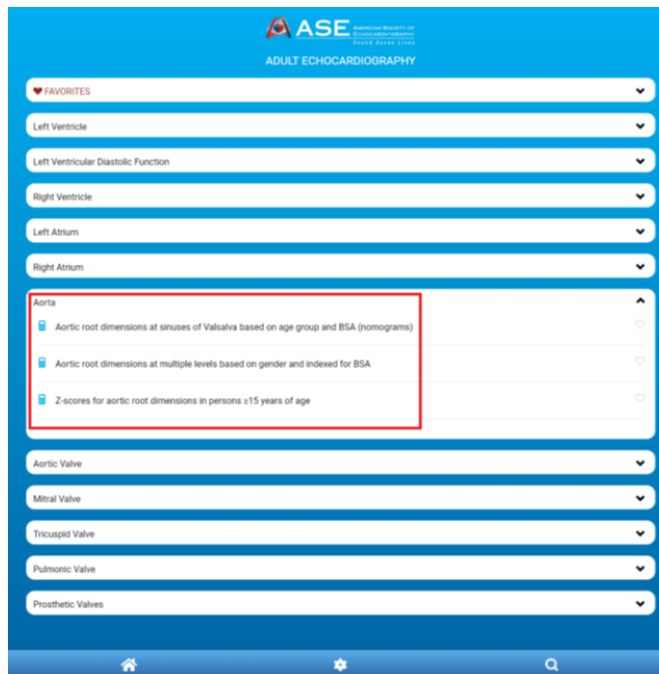


Figure 2 Transthoracic echocardiogram in the parasternal long-axis view (zoomed on aortic root and ascending aorta) illustrating measurement of the aortic root diameter at sinus of Valsalva level at end-diastole using the leading edge-to-leading-edge method. asc Ao, Ascending aorta; LVOT, left ventricular outflow tract.



@EGarciaSayan:

▲ Sensibilidad de ETT en ASV >90%

▲ Identificar ubicación, tamaño, relación y compresión de estructuras adyacentes, flujo anormal con Doppler color

▲ Descartar insuficiencia aórtica & lesiones asociadas

▲ ETE para visualizar aorta y coronarias

#ASEchoJC

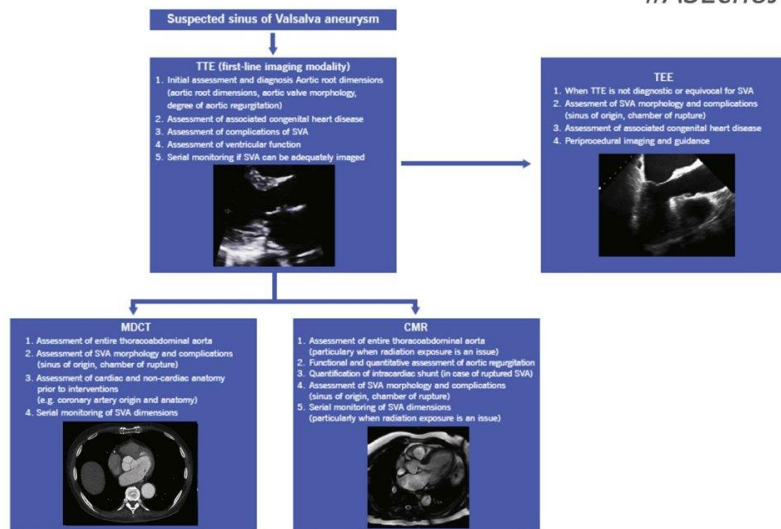
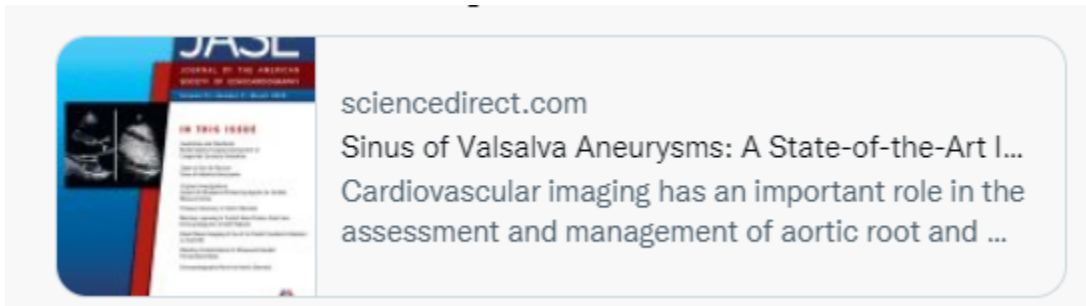


Image from: J Am Soc Echocardiogr. 2020 Mar;33(3):295-312.

@EGarciaSayan:

- ▲ Imágenes adicionales en ASV incluye uso de #YesCCT & #WhyCMR
- ▲ Delinear anatomía y relación con estructuras adyacentes
- ▲ Asistir con plan operativo
- ▲ Ver excelente review de imagen multimodal en ASV en @JournalASEcho: <https://t.co/mSMRkySI5Y>



@nilda_zavaleta: En el caso de la ruptura del ADSV, la evaluación ecocardiográfica con Doppler color revela un flujo continuo en sístole y diástole, aleteo de la válvula tricúspide a medida que el chorro de color se mueve desde la aorta a las cavidades derechas del corazón

@nilda_zavaleta: Es importante definir la localización del seno comprometido y de la porción sinotubular de la aorta, la coexistencia de aorta bicúspide, el compromiso del sistema de conducción,

@nilda_zavaleta: la presencia y tipo de cortocircuitos, la severidad y mecanismo de insuficiencia aórtica y la cercanía de los ostia coronarios

@nilda_zavaleta: Tanto los ADSV rotos como los no rotos pueden complicarse por insuficiencia aórtica, que ocurre en 30-50% de los pacientes. La resonancia cardíaca con secuenciación multiplanar ha permitido la evaluación de cortocircuitos intracardíacos en ADSV rotos


@nilda_zavaleta: Nuevos generadores de imágenes de TC cardíaca permiten obtener imágenes diagnósticas precisas incluso de ADSV pequeños en un entorno dinámico. De acuerdo con las guías de enfermedades de la aorta torácica de la AHA 2010, la reparación quirúrgica debe considerarse en

@nilda_zavaleta: aquellos con aneurismas >5.5 cm, >5 cm en aquellos con válvula bicúspide, >4,5 cm en el contexto de enfermedad del tejido conectivo, o una tasa de crecimiento de >0.5 cm/año

@nilda_zavaleta: Se recomienda la intervención quirúrgica para un ADSV roto y/o un ADSV con anomalías intracardíacas asociadas, como CIV o insuficiencia aórtica. También se debe considerar la reparación quirúrgica en un ADSV no roto, pero sintomático o dilatado

Question 10:

Question 10 #ASEchoJC

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What does the surgeon want to know from the #EchoFirst images about the SOVA?

11/15/22 @SuntharKarMD @nilda_zavaleta @VLSorrellImages @boegel_kelly @EGarciaSayan @iamritu

A10 Notable responses

@EGarciaSayan: surgical indications in SOVA

- ▲ Ruptured: early intervention (↑ 1-year💀)
- ▲ Symptomatic, non-ruptured: Malignant arrhythmias, endocarditis, coronary or RVOT obstruction
- ▲ Asymptomatic: Rapid expansion or worsening AR?
- ▲ Percutaneous closure emerging in selected cases

@nilda_zavaleta: The surgeon wants to know the diameter of the SOVA, the competence and severity of pathological changes of the aortic valve, the proper position of the SOVA rupture and coronary ostia

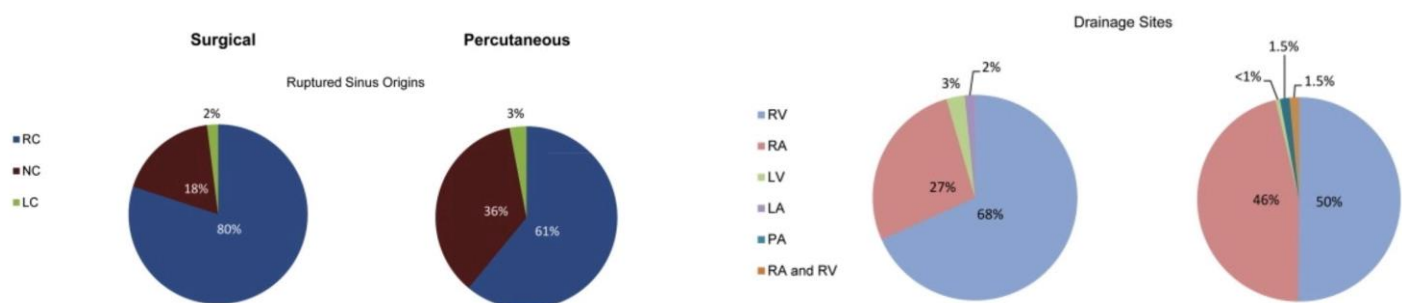
@nilda_zavaleta: The presence and type of the associated structural abnormalities, myocardial involvement (myocardial characterization and quantification of left ventricular mechanics) and associated RV involvement

@iamritu: Surgical approach based on ruptured chamber, assoc lesions

Percutaneous closure an alternative w isolated RSA but Sx for those w multiple cardiac lesions

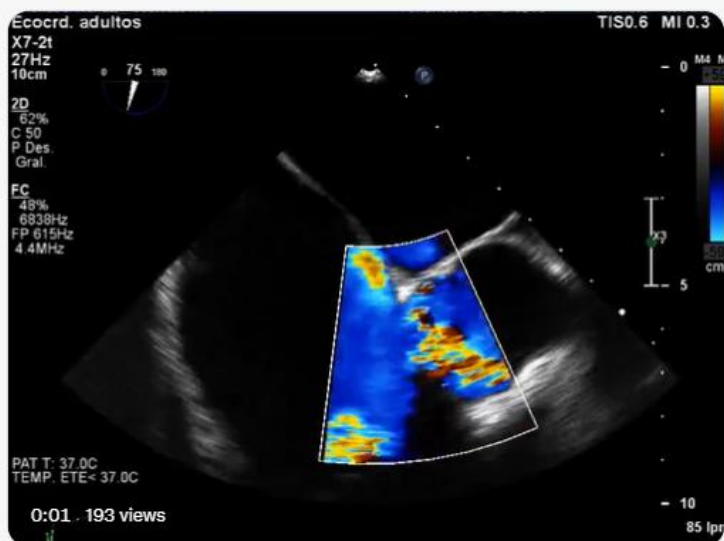
closure depends on aneurysm size, relationship of aneurysm w adjacent structures, shape

<http://bit.ly/3X0npTT>



@VLSorrellImages: Surgeons like seeing pre-op images like this:

<https://twitter.com/i/status/1592697896591163392>



@boegel_kelly: Keep these things in mind when you are scanning a patient with SOVA

@EGarciaSayan: indicaciones de cirugía en ASV

- ▲ Ruptura: Intervención temprana (👆💀)
- ▲ Síntomas sin ruptura: Arritmias malignas, endocarditis, obstrucción coronaria o del tracto VD
- ▲ Asintomático: ¿Expansión rápida insuficiencia Ao?
- ▲ Considerar cierre percutáneo en casos selectos

@nilda_zavaleta: El cirujano quiere saber el diámetro del ADSV, la funcionalidad y gravedad, de los cambios patológicos de la válvula aórtica, localización precisa del ADSV roto y los ostia coronarios

@nilda_zavaleta: La presencia y el tipo de anomalías estructurales asociadas, el compromiso del miocardio (caracterización miocárdica y cuantificación de la mecánica ventricular izquierda) y el compromiso del VD

@EGarciaSayan: And that's all for tonight's #ASEchoJC, which we are dedicating to the memory of our dear colleague and friend, Greg Tatum 🙏

If you missed live tweets, catch up on the discussion later by searching #ASEchoJC.

👉 Claim CME: <https://bit.ly/3Ai8BGe>

👉 See you again in a few weeks <https://twitter.com/i/status/1592699079016210432>

Thank you.

GIF