Peer Review File

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Reviewer A

1. The case that has been described by the authors is very interesting. Would the authors be able to comment on the nuchal thickness in the patient prenatally?

Reply 1: Thank you for your interest in our case. Regarding nuchal thickness, we have added additional information to the manuscript to confirm that the patient's prenatal nuchal thickness was within normal limits.

Changes in the text: Page 8, line 107

2. Why did the patient present for a fetal echocardiogram at 31 weeks gestation? Was this echogenic structure seen on prior prenatal ultrasounds/fetal echocardiograms? What was the progression of the RV size and tricuspid and pulmonary valve sizes during the antenatal period?

Reply 2: Thank you for your questions. The patient was initially referred to our center at 31 weeks of gestation for targeted fetal echocardiography due to findings of an “abnormal tricuspid valve and right heart development” observed during an ultrasound examination at 24 weeks of gestation. Subsequent follow-up imaging at 37 weeks at their local hospital revealed no significant change in cardiac findings compared with the echocardiogram performed at our center. Additionally, no other abnormalities in fetal development were noted. The patient was then monitored by routine ultrasound assessments thereafter.

Changes in the text: Page 8, line 101-117

3. The authors report that a baby was born at a different center. Was this the same baby that had been followed prenatally? Could the authors verify this?

Reply 3: Thank you for pointing out the need to clarify this matter. The baby born at a different center was the same baby that had been followed prenatally. We have revised the manuscript to make this clear.

Changes in the text: Page 8, line 118

4. Can the authors provide the degree of obstruction across the tricuspid valve as the membrane became bigger? It appears that there were at least 2 to 3 echocardiograms performed postnatally. Was the obstruction to inflow due to the restriction in the membrane in the right atrium or due to tricuspid valve hypoplasia or a combination of these factors?

Reply 4: In response to your query, we conducted three echocardiographic evaluations, and the degree of obstruction across the tricuspid valve (TV) was primarily assessed through echocardiographic parameters, including narrowing of the tricuspid inflow to 3 mm and a maximal Doppler velocity of 1.4 m/s. These values were consistent across two subsequent bedside echocardiograms. However, the last two bedside assessments focused primarily on the patient's cardiac function, as well as the patency of the foramen ovale and arterial duct. We observed no significant increase in size of the membranous structure in the postnatal period based on the available echocardiograms, suggesting that

the obstruction remained relatively stable. Closer examination of the relationship between the membranous structure and the TV showed that the accelerated jet of blood flow primarily originated from the space between the membrane and the TV. We believe the main problem with the TV blood flow was due to the obstruction caused by the membranous structure, rather than to an inherent developmental anomaly of the TV itself. Accordingly, normal blood flow through the TV was restored after surgical removal of the membranous structure, further supporting this point. This also suggests that the developmental issues with the right ventricle (RV) and TV were likely due to this obstruction, rather than to an intrinsic problem with the TV.
We have added this information to the revised manuscript to provide a more comprehensive description and explanation, and would welcome any further valuable feedback.
Changes in the text: Page 9, line 126-129

5. Can the authors comment on the pulmonary valve stenosis gradient postnatally on the serial echocardiograms?
Reply 5: Thank you for your pertinent question. In our study, we did not observe any significant anomalies in pulmonary valve morphology, with no evidence of an abnormal number of pulmonary valve leaflets, leaflet thickening, or increased echogenicity; however, the pulmonary valve annulus appeared to be slightly narrow (Z score, −2.5) during outpatient follow-up on the 10th postnatal day. We found no evidence of significant forward blood flow acceleration (<1.5 m/s). Subsequent bedside ultrasound examinations also failed to show any significant accelerated flow signals. We believe that these findings may be related to reduced blood flow secondary to right ventricular inflow tract obstruction, rather than to primary pulmonary valve stenosis. The blood flow and inner diameter normalized as the obstruction resolved. We have added a section to the revised manuscript to elaborate on the dimensions of the pulmonary valve annulus.
Changes in the text: Page 9, line 129-132

6. Can the authors comment on why the baby developed worsened desaturations on day of life 25 as opposed to within the first 1 to 2 weeks after birth?
Reply 6: We attributed this delayed onset of worsened desaturations to a potential concurrent pulmonary infection. This additional complication could have contributed to the deteriorating oxygen saturation levels seen at that time, compared with the earlier, more-stable period. We have clarified this point in the revised manuscript to enhance understanding of the clinical course of this case.
Changes in the text: Page 10, line 145-148

Reviewer B
Overall, very good case report and review. Please see our review below,
Page 1:
Abstract: Background: “depending upon the course of the abnormality”□ depending on the severity of the abnormality
Reply 1: We have modified the text as advised.

Changes in the text: Page 4, line 53-54.

Case Description: “newborn who was followed from 31 weeks gestation” newborn who was followed since fetal life starting 31 weeks of gestation”.
Reply 2: We have modified the text as advised.
Changes in the text: Page 4, line 57.

Body of manuscript by lines:
Line 102: “fluctuating hyperechogenic membrane” “mobile hyperechogenic membrane”
Reply 3: We have modified the text as advised.
Changes in the text: Page 8, line 113.

Line 103: and Eustachian valve (EV) and prominent Eustachian valve
Reply 4: We have modified the text as advised.
Changes in the text: Page 8, line 114.

Line 108: … ductus arteriosus (1.0 mm). Was there really bidirectional flow at the PDA? That’s a bit unusual, since typically with hypoplastic right heart lesions you would expect more ductal dependency and purely left to right flow. Can you provide more confirmatory images of this, and explain why this baby displaced bidirectional shunting which would imply pulmonary HTN at 10 days of age?
Reply 5: Thank you for your insightful comments and for pointing out this important issue. We apologize for the confusion. To clarify, we intended to describe bidirectional shunting at the PFO, not at the PDA. The PDA in this case actually demonstrated left-to-right shunting. We realize that our original wording may have led to some ambiguity, and we appreciate the opportunity to rectify this.
Changes in the text: Page 8, line 121.

Line 109: Can you please provide details about the degree of hypoplasia involving the right side? What was the tricuspid valve z-score? What about the right ventricle, other than what was provided for the RVOT dimension and z-score? Was the RV apex forming? Was it tripartite or bipartite? This is important since later in report you will describe that the right ventricle normalized 5months after surgery, raising the question of whether the RV was more “underfilled” rather than truly hypoplastic.
Reply 5: Thank you for your insightful comments. The three components of the RV (inlet, apical trabecular, and outlet) were all present but demonstrated moderate hypoplasia. We have added the following measurements to provide a more comprehensive understanding of the right-sided hypoplasia: basal diameter, 8.2 mm (Z-score, −3.4), long-axis diameter, 13.8 mm (Z-score, −4.6), and outflow tract diameter, 6.8 mm (Z-score, −4.4). The TV was hypoplastic but morphologically normal, with an annulus diameter of 7.2 mm (Z-score, −2.5). The pulmonary valve annulus measured 6.9 mm (Z-score, −2.3) with no abnormalities, and the pulmonary artery was 6.7 mm (Z-score, −2.7) with no evidence of flow acceleration.
Additionally, we would like to correct the follow-up time: the available echocardiographic

data were measured at 3 months of follow-up, at which point the size of the right heart had essentially normalized.
Changes in the text: Page 9, line 122-125;129-132. Page 11, line 168.

Line 109: what was the z-score of the pulmonary valve? You mentioned it was normal, but with a hypoplastic RVOT and a low normal MPA you would expect a hypoplastic pulmonary annulus.
Reply 5: Thank you for bringing this to our attention. We have added the Z-score of the pulmonary valve to the revised manuscript for clarification. The pulmonary valve annulus was 6.9 mm, with a Z-score of −2.3.
Changes in the text: Page 9, line 129-132.

Line 111: “A redundant soft membrane was observed fluctuating”. Was it really soft? In utero it was described as hyperechogenic. Also, fluctuating is again probably not the strongest term for this. “displaying mobility” or some other term may be more appropriate.
Reply x: Thank you for your detailed observation concerning the description of the membrane's texture and mobility. Upon reviewing the ultrasonographic images, we agree that the term “hyperechogenic” may not be the most accurate description for the *in utero* findings. We have described the membrane as “echogenic” during the prenatal period in the revised manuscript. We also agree that “fluctuating” might not provide the best description of the membrane's movement, and have therefore replaced “fluctuating” with “mobile” in the revised manuscript, to offer a more precise description.
Changes in the text: Page 8, line 113. Page 9, line 126.

Line 112-113: “the RA and was adhered to the atrial septum, and it reached the FO” Is there a better description?
Reply x: Thank you for your insightful comment. We have revised this description to improve the clarity. The manuscript now specifies that “a mobile, echogenic membrane was observed in the RA near the superior vena cava (SVC), which prolapsed through the TV into the RV….”. We hope that this modification provides a clearer understanding of the observed anatomical details.
Changes in the text: Page 9, line 126-127.

Line 113: what does flow shunting through the tricuspid valve mean???
Reply x: Thank you for bringing this to our attention. The phrase “flow shunting through the tricuspid valve” was indeed a typographical error on our part. We have rectified this in the revised manuscript.
Changes in the text: Page 9, line 126-129.

Line 125: please comment on direction of flow across PFO and PDA.
Reply x: Thank you for highlighting this important aspect. We apologize for any confusion regarding our original description. We have revised the manuscript to clarify that the flow across the PFO was bidirectional, while the flow across the PDA was predominantly left to right. We believe that specifying these details will allow a more comprehensive

understanding of the hemodynamics involved.
Changes in the text: Page 10, line 148-149.

Line 127: “and pulmonary blood flow”□ and provide an additional source of pulmonary blood flow.
Reply x: Thank you for your suggestion. The addition of "and provide an additional source of pulmonary blood flow" adds important context and specificity to the phrase "and pulmonary blood flow."
Changes in the text: Page 10, line 153-154.

Line 130: “and the effective response failed to be stable”: redundant, you can just delete this phrase.
Reply x: Thank you for pointing out the redundancy. We have removed the phrase “and the effective response failed to be stable” from the manuscript.
Changes in the text: Page 10, line 157.

Line 133: “A fibrous fenestrated membrane”□ “Intraoperatively, a fibrous fenestrated membrane”.
Reply x: Thank you for this specific guidance. We have amended the sentence as suggested.
Changes in the text: Page 11, line 166.

Line 135: please describe attachments to the atrial septum, and if possible the size of effective orifice opening into the right atrium.
Reply x: Thank you for raising this issue. We apologize for not making this clear. To clarify, we used a minimally invasive approach during the surgical procedure. Unfortunately, the exact size of the effective orifice opening was not measured intraoperatively; however, the dimension was assessed using echocardiography and has been added to the revised manuscript for further clarification. During the surgery, the attachments to the atrial septum were further examined via transesophageal echocardiography, which showed that the membrane originated from the anterior margin of the SVC in the RA and attached to the TV. We have amended the manuscript to include this information.
Changes in the text: Page 9, line 126-129. Page 10, line 161-163.

Line 136: what does minimally invasive approach mean? Please describe what makes your surgical approach minimally invasive.
Reply x: We appreciate the opportunity to clarify this issue. Our minimally invasive approach referred to the use of a smaller midline incision and entry through the right fourth intercostal space, thus reducing trauma to the chest wall and minimizing blood loss. Additionally, we utilized a 9F delivery sheath and 2F biopsy forceps to capture and remove the network, all of which were guided by transesophageal echocardiography. We have expanded on this in the revised manuscript.
Changes in the text: Page 10-11, line 163-166.

Line 139: again, please provide measurements of TV and RV, and description of RV (apex, forming, tripartite, etc).
Reply x: Thank you for your insightful suggestion. We agree that providing measurements and a thorough description of the TV and RV would add valuable context to our study. We have included specifics in the revised manuscript, including the dimensions of the TV and RV, and a detailed description of the RV.
Changes in the text: Page 11, line 169-172.

Lines 162-164: “We collected a series of PRVV cases from prenatal to postnatal periods and discussed the disease course, clinical presentation, treatment methods, and prognosis. Compared to single-case studies in the literature, our research, involving more cases, could provide additional information and data”. This line is confusing and misleading. It leads the reader to think you may have collected cases at your institution, and it looks like you collected cases from literature review. Perhaps it is better to say: “We identified similar cases reported in the literature, and summarized the reported prenatal echocardiographic features (Table 1), and echocardiographic and clinical features as well as surgical repair of other cases (Table 2).”
Reply x: Thank you for pointing out the ambiguity. We have revised this section for clarity.
Changes in the text: Page 12, line 201-203.

Line 173: “The CN is considered a variant of the EV” The Chiari network is a distinct structure that is different from, though related to, the eustachean valve.
Reply x: Thank you for your valuable input. We have updated our manuscript to specify: "The CN is an extended, fenestrated, echogenic structure that originates from either the Thebesian valve or EV and attaches to the RA wall or interatrial septum." We hope this provides a more accurate representation of the anatomical relationship between the Chiari network and the Eustachian valve.
Changes in the text: Page 13, line 214-216.

Line 176 to 178: “Additionally, the cor triatriatum dexter (CTD) originates from the crista terminalis and is characterized by the partitioning or septation of the RA to form a triatrial heart.” Is PRVV really different from CTD? If so, please educate the reader more on how you make that distinction echocardiographically in this section. You do so later on line 233, but better to do it in this section since you already alluded to the comparison here. And when you make that distinction based on the appearance, is that your own personal opinion? Or is that an agreed upon distinction? There are many clinicians who think of PRVV and CTD as being the same entity, or on the spectrum of basically the same disease.
Reply x: Thank you for your insightful comments.
CTD and other anatomic variants of PRVV share a common embryological origin in the right sinus venosus valve, but their developmental pathways diverge. Specifically, CTD is characterized by additional attachments to the atrial septum giving the appearance of a divided atrium, while other anatomic variants of PRVV typically present with a less-extensively developed membranous structure within the RA. Echocardiographically, they exhibit different characteristics and origins. CTD originates from the crista terminalis and

involves partitioning of the right atrium, whereas other PRVV variants consist of a membrane or network of fibers attached to the RA or interatrial septum. When distinguishing these entities based on their appearance, the observations are supported by current medical literature and case reviews.
We appreciate your suggestion to elaborate on this distinction and have revised the text accordingly.
Changes in the text: Page 13, line 218-222.

Lines 179-214: too wordy, detailed, repetitive, and redundant. As a reader at this point I started to lose focus and got the feeling that I was no longer learning anything new. Please make this section more concise and less repetitive if possible.
Reply x: Thank you for your valuable feedback. We have revised this section to make it more concise, while retaining the essential details.
Changes in the text: Page 13-14, line 223-252.