

BIDIRECTIONAL GLENNSHUNT OPERATION IN A PATIENT WITH CANTRELL SYNDROME AND LEFT VENTRICLE DIVERTICULUM

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Cantrell Syndrome (CS) is defined as a combination 5 major anomalies, also known as Pentalogy, which involves a midline anterior abdominal wall defect, a distal sternal cleft, a defect of the anterior diaphragm, and a defect of the apical pericardium with pericardio-peritoneal communication, as well as intracardiac anomalies. This anomaly is a rare entity and is encountered in 5.5 per 1 million live births.¹ The severity of the syndrome is dependent on the associated cardiac defects. In addition to transthoracic echocardiography, computed tomography angiography (CTA) defines the co-existing defects and delineates the complete anatomy (Figure 1).² The protrusive cardiac segment ranges from isolated left ventricular diverticulum (LVD) to total ectopia cordis. About 70% of the reported LVD cases have CS.³

We present a case of a 5 month-old boy weighing 4.8kg's who had right ventricle hypoplasia, mesocardia, crisscross ventricles, and atrioventricular septal defect. The patient had previously undergone a pulmonary artery banding operation with an upper mini sternotomy in the neonatal period. Although early surgical intervention is advocated even in asymptomatic patients considering the potentially detrimental effects such as embolization, tachyarrhythmias, spontaneous or traumatic rupture, endocarditis, and sudden cardiac death, the excision of the LVD was delayed to a second stage due to the critical hemodynamic status at presentation.

The surgical excision of LVD requires several precautions. The skin incision should be limited until the edge of the sternum whenever possible. Due to the segment with

abdominal wall defect being considerably thin, the healing in this segment may be difficult. Furthermore, cardiopulmonary bypass has to be readily available in case of emergency. The LVD should be thoroughly inspected to rule out existing coronary vessels adjacent to the diverticulum. Furthermore, during these manipulations, hemodynamic stability is essential and requires advanced anesthesiological management. When the LVD is totally exposed, following the insertion of the clamp in the diverticulum's neck, the surgeon has to detect possible ventricular arrhythmia and blood pressure changes that may necessitate clamp adjustment or institution of cardiopulmonary bypass. If the patient remains hemodynamically stable, the LVD can be resected and sutured using polypropylene interrupted stitches with pledgets (Figure 2), (Video 1).

In our case, we then performed an off-pump bidirectional Glenn Shunt (BDG) and division of the main pulmonary artery. However, an extrapulmonary artery arising from the upper surface of the right pulmonary artery associated with Bronchus Suis was observed. However, this condition did not pose any obstacle to the anastomosis of the BDG.

In conclusion, we state that early operation is indicated in LVD without hesitation considering the life-threatening side effects. The CTA provides important details regarding the defect to confirm and reveal the exact anatomy. The removal of the LVD can be performed with or without CPB. Yet, all precautions must be taken and the surgeons have to always keep in mind that every step of the surgery requires great care.

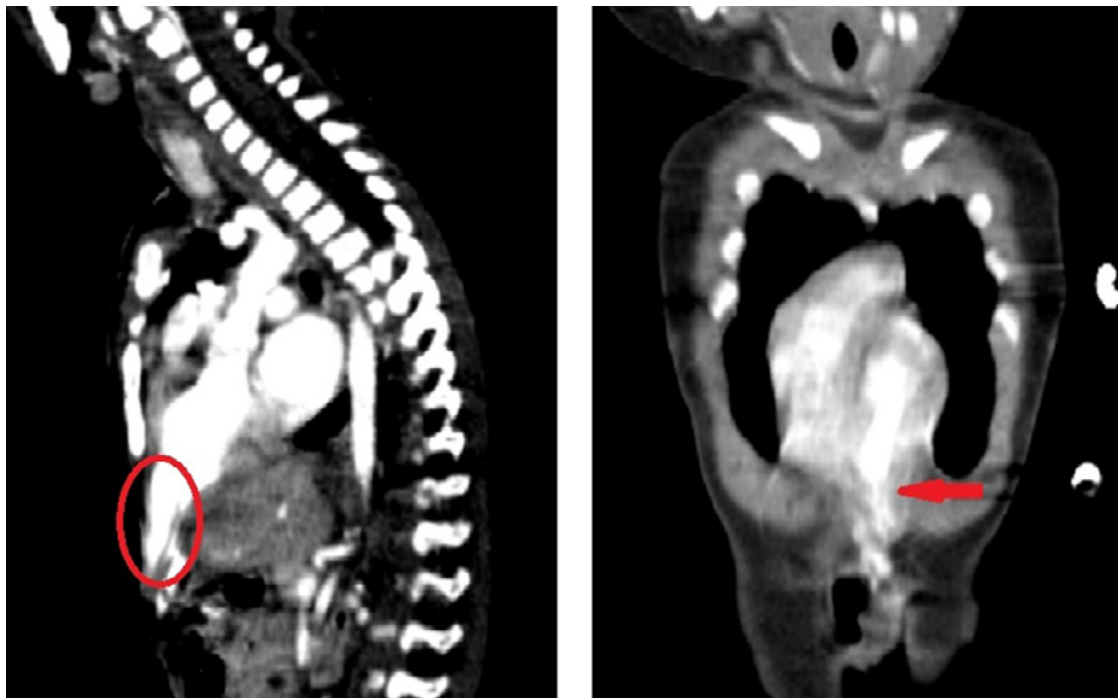


Figure 1 *The computed tomography images of the left ventricle diverticulum*



Figure 2 *The images of the resected diverticulum. 10F hegar bugie was able to be inserted to the lumen of the diverticulum.*

Patient Consent for Publication: A written informed consent was obtained from parents of the patient.

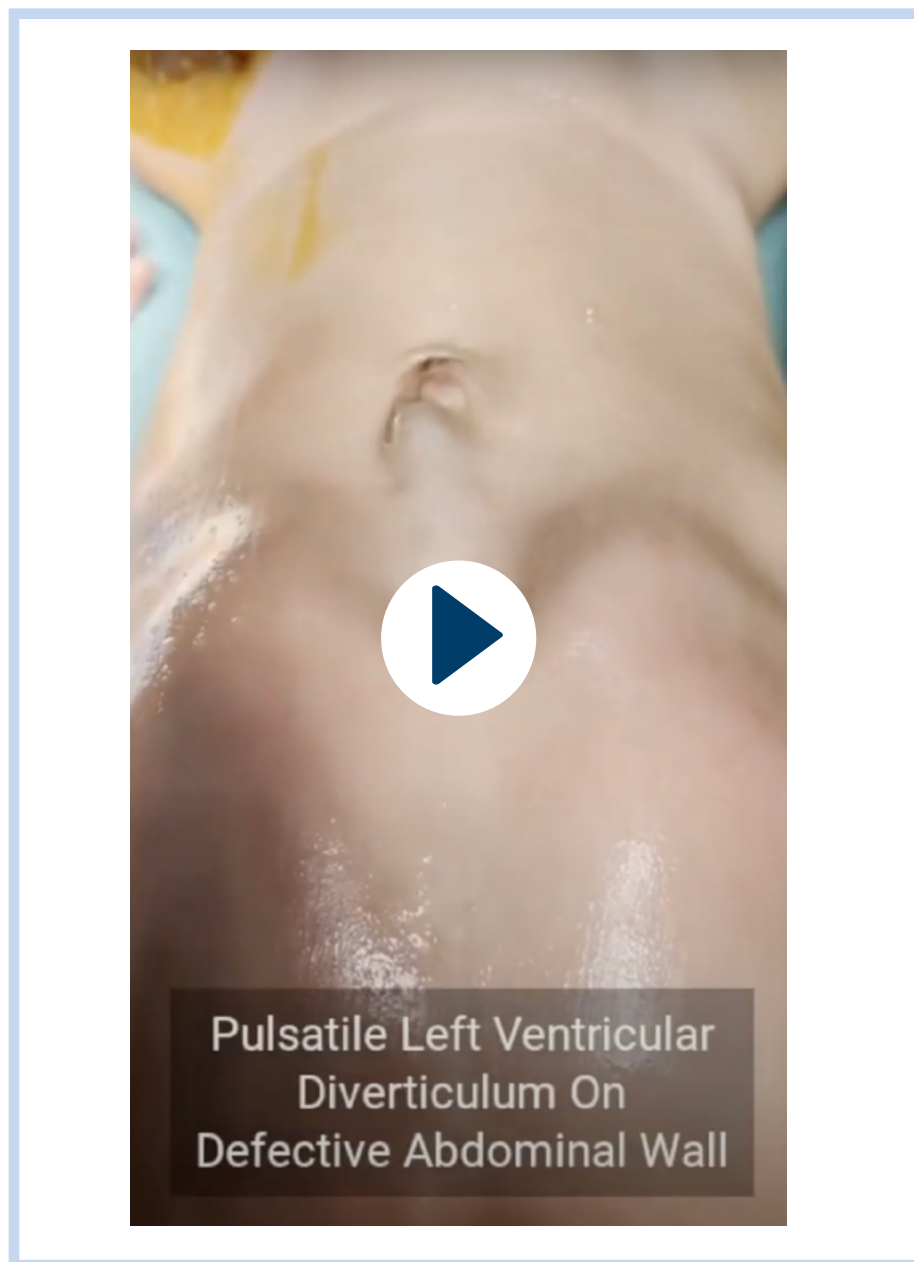
Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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Video 1

Part 1: The preoperative image. The pulsatile LVD is apparent on the defective abdominal wall
 Part 2: The image before sternomy. The skin incision is limited on the edge of the sternum
 Part 3: The LVD is suspended by silastic tape
 Part 4: The LVD is totally exposed, no coronary artery on the surface on the LVD
 Part 5: The extra right pulmonary artery branch associated with bronchus suis
 Part 6: Postoperative image. The stumps of the LVD, main pulmonary artery and the anastomosis of the BDG are shown respectively.

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