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Reconstruction of Congenital Sternal Clefts: Surgical Experience and Literature Review

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Background : Sternal cleft is a rare anomaly with a reported incidence of 1:100,000 cases per live births. Surgical intervention represents a crucial factor altering the overall patient prognosis, since they are at high risk of impaired oxygenation, as well as multiple chest infections. Herein, we are reporting our experience of surgical management of such rare cases, alerting plastic surgeons to their possibly crucial role in the reconstructive team.

Methods: A retrospective chart review of 2 cases presenting with chest wall defects. All perioperative data were collected and presented.

Results: Two patients with sternal clefts of variable degrees were managed. The first was an 18-month-old boy with partial inferior sternal cleft, who was otherwise asymptomatic. The patient underwent reconstruction at the same age using autologous rib graft and pectoralis major flaps due to ectopia cordis that was putting the patient at higher risk for cardiac trauma. The second patient was a 3-month-old girl having a V-shaped partial superior cleft with lung herniation. Surgical reconstruction was applied due to difficulty in weaning the patient off of ventilator support. Initially, reconstruction was applied with SurgiMend dermal matrix, but this was complicated by chest retraction and high oxygen requirement. Definitive reconstruction was later applied with allogeneic bone graft and pectoralis major flaps.

Conclusions: Meticulous patient assessment and screening for associated anomalies are crucial. Surgical intervention is warranted at an early age. The use of acellular dermal matrix products in the reconstruction is of interest, but should be approached with caution. *(Plast Reconstr Surg Glob Open 2017;5:e1567; doi: 10.1097/GOX.0000000000001567; Published online 20 November 2017.)*

INTRODUCTION

Anterior chest wall malformations include various musculoskeletal abnormalities like sternal clefts, pectus excavatum, pectus carinatum.[1] Sternal cleft is a rare anomaly with a reported incidence of 1:100,000 cases per live births and represents less than 1% of all chest wall congenital deformities.[2] Sternal cleft was first identified in 1974 and later on truly identified pathologically by E.A. Groux in 1858.[3] The embryonic origin of the sternum was first linked to different primordial origins related to ribs or the anterior pectoral girdle; this, however, was challenged, whereby an independent pair of mesenchymal condensations in the somatopleuric mesoderm was identified as the true origin of the sternum's development.[4] The sternum formation starts at the sixth week of gestation, with the lateral mesodermal plates advancing ventrally to create 2 parallel mesenchymal strips. During the seventh week of gestation, these strips start to merge cranio-caudally, creating the sternal body and part of the manubrium with complete fusion in the 10th week as cartilaginous primordia. Multiple ossification centers then develop in a cranio-caudal sequence; however, fusion of those centers is in a reverse direction.[3–5] Many efforts were made to classify sternal clefts. Shamberger and Welch[6] have suggested 4 categories of clefts that involve cervical ectopia cordis, thoracic ectopia cordis, thoracoabdominal ectopia cordis, and sternal cleft or bifid sternum. Later description was made based on the extent of the cleft process, as either partial or complete sternal cleft, where the partial malformations affect either the superior or inferior sides of the sternum.[7] Superior partial sternal clefts represent the

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commonest malformation, with a characteristic U-shaped cleft reaching up to the fourth costal cartilage.[8] Patients with sternal clefts are at high risk for respiratory tract infection and lack of protection to vital mediastinal structure.[3] Surgical correction of sternal clefts represents a crucial modality of the treatment and was linked to the patient's age, being a decisive factor in determining the possibility of primary approximation. This was recommended during the neonatal period, during which chest wall elasticity will permit primary closure.[9,10] Beyond this age frame, and presumably after 3 months of age, the chest wall becomes rigid and more complicated surgical techniques would be warranted.[11] Among techniques described are sliding chondrotomies, partial or total thymectomy, clavicle dislocation, bone or cartilage graft interposition, and muscle flap interposition.[3,11,12] The use of different prosthetic as well as biologic mesh grafts was also utilized to aid in the closure of such defects with variable outcomes.[13,14]

Herein, we are reporting our experience in the surgical management of such rare cases of sternal abnormalities and discuss the outcomes obtained. Distinct patient presentation was associated with different treatment plan. The essential role of plastic surgeon being familiar with variable autologous as well as alloplastic graft options was shown, being a crucial member in the reconstructive team.

PATIENTS' PRESENTATION AND MANAGEMENT

Patient 1

An 18-month-old baby boy, product of a full-term twin pregnancy with a birth weight of 2.4 kg and having an uneventful antenatal care, was referred from the thoracic surgery section with the complaint of lower midline chest wall bulge for surgical correction. History was significant for ventricular septal defect, and moderate tricuspid valve regurgitation, in which the patient had received a cardiac correction surgery. The patient was fitted with a protective garment to prevent any trauma to the underlying heart; he was otherwise asymptomatic. Preoperative computed tomography (CT) scan showed partial inferior sternal cleft with associated ectopia cordis, in which the left ventricle was lying immediately under thin cutaneous coverage. The remainder of the physical examination was otherwise normal. Operative intervention to reconstruct the defect was undertaken. The procedure started with marking the costal margin, followed by skin infiltration with diluted infiltration solution (1 ml of 1% lidocaine with epinephrine mixed in 20 ml of normal saline) to allow for hydro-dissection and a bloodless field in direct communication with anesthesia and thoracic surgery teams (Figs. 1, 2). Dissection followed, in which both the pectoralis major and the rectus abdominis muscles were raised as single units bilaterally. The right seventh rib was harvested and was divided into sections and secured with polydioxanone absorbable suture to the cleft edges bilaterally bridging the defect over the heart. The muscles were then advanced and approximated to the midline covering the rib graft with the use of absorbable sutures followed by skin closure. Patient had an uneventful postoperative course and was discharged home on the third day postoperatively. The child was doing well with uncomplicated course in 6 months follow-ups.

Patient 2

A 3-month-old baby girl, a product of full-term pregnancy with a birth weight of 3 kg with uncomplicated pregnancy. The patient was referred from the pediatric surgery department with the complaint of a superior midline chest wall bulge with paradoxical wall movement for surgical reconstruction. The patient was initially admitted to the pediatric intensive care unit (PICU) after acquiring respiratory distress and lung collapse, complicated with respiratory tract infection and was on mechanical ventilator with difficult extubation. After being stabilized, CT scan was obtained and showed a V-shaped partial superior sternal cleft with separation of the ossification center of the manubrium and the upper part of the body of sternum. The distance between the manubrium sides was 4 cm with partial herniation of the left lung and upper mediastinal structures through the sternal cleft. The heart was normal in location (Figs. 3, 4). Remaining examination and investigations were normal. The patient was taken to the operating theatre, where the procedure started with skin infiltration as described in the previous case. Dissection was then carried out to raise the pectoralis major muscle bilaterally as advancement flaps. The cleft margins were then dissected while protecting the underlying mediastinal structures. Intraoperative assessment showed difficulty in harvesting a rib for possible autogenous graft. The decision was made to utilize a 4-mm thick acellular dermal matrix (ADM) of fetal bovine origin (SurgiMend) as a mean to bridge and stabilize the cleft. The mesh was then secured to the cleft edges with the use of polydioxanone absorbable sutures (Figs. 3, 4). Bilateral pectoralis major muscles were then advanced to the midline and secured over the mesh. This was followed by skin closure. Postoperatively, the patient was kept for observation in the PICU. Following extubation on the fourth day postoperatively, the patient started to develop inward recoil of the chest wall at the cleft site with increasing oxygen requirements. She was maintained on high flow oxygen through nasal cannula. The patient remained in PICU with no change in her condition for about 8 weeks. The patient was then taken to the operating room during which definitive skeletal stabilization was applied. Utilizing the same incision, the skin and muscles were dissected to reach the SurgiMend level and cleft edges. Allogeneic bone graft obtained from a local bone bank was then cut and fashioned to fill the defect. The graft was then secured to the cleft edges with fixation wires (Figs. 3, 4). Postoperatively, the patient was doing well with uncomplicated course in 6 months follow-ups.

DISCUSSION

Sternal clefts are among the rarest congenital anomalies that affect the developing anterior chest wall, with less than 1% among all chest wall deformities.[2] Many factors were linked to the development of this disease entity, such as methyl-cobalamin or riboflavin deficiency, alcohol in-

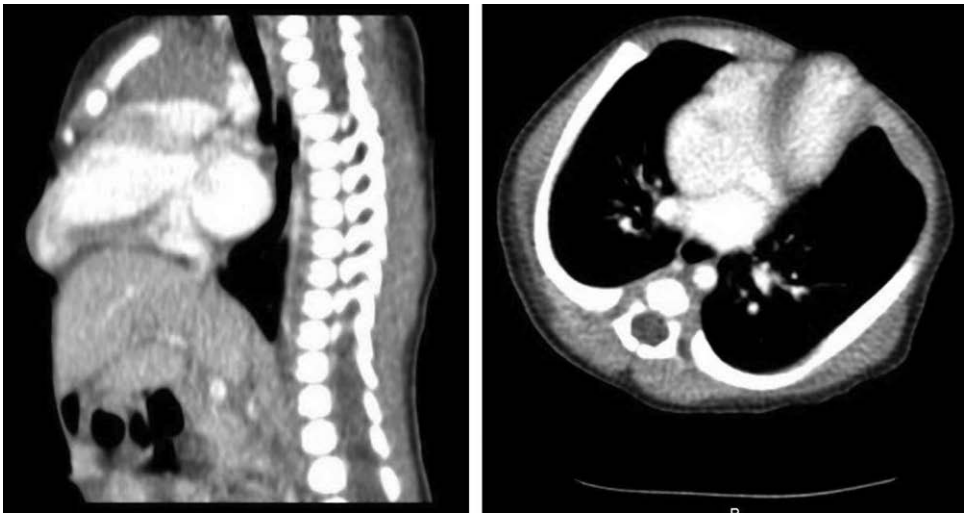

Fig. 1. Axial section of preoperative CT scan. Shows partial inferior sternal cleft and associated ectopia cordis under thin cutaneous coverage.

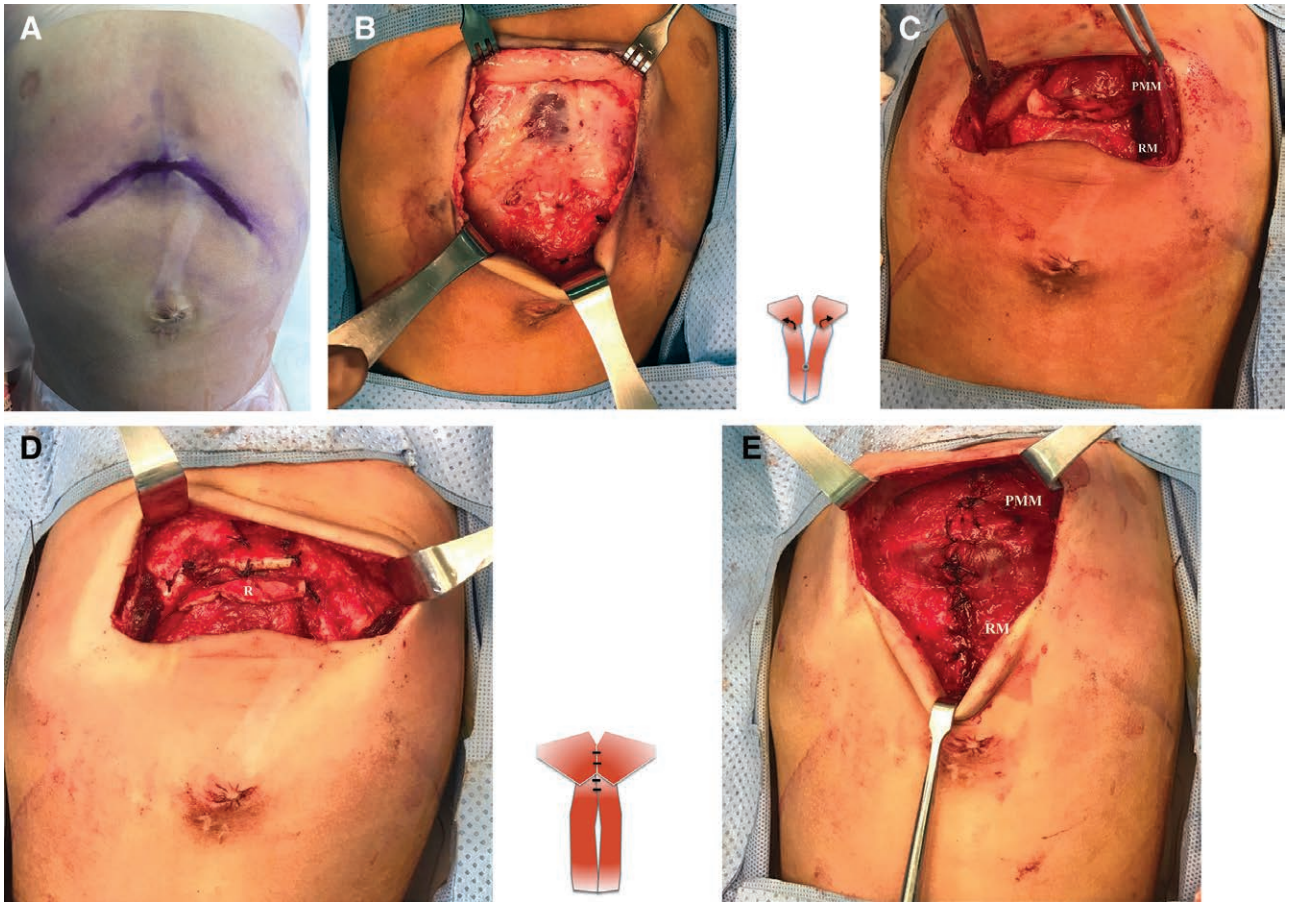

Fig. 2. Operative planning and reconstruction. Horizontal incision line followed by raising cutaneous flaps (A and B), followed by raising bilateral pectoralis major and rectus abdominis muscles as single units bilaterally (C); rib graft was then harvested and secured to the cleft margins after being cut into several segments (D). The muscles were then advanced and mobilized to the midline to cover the rib grafts (E). PMM, pectoralis major muscle; RM, rectus abdominis muscle; R, rib.

take and HOX b gene anomalies.[7] The disease in general has female predominance.[12] When a patient is identified as having a sternal cleft, meticulous screening and patient assessment are warranted to rule out any associated anomalies that might alter the overall surgical prognosis. The associated conditions described include hemangiomas or other features of Posterior fossa anomalies, Hemangiomas, Arterial anomalies, Cardiac defects, Eye abnormalities, Sternal cleft

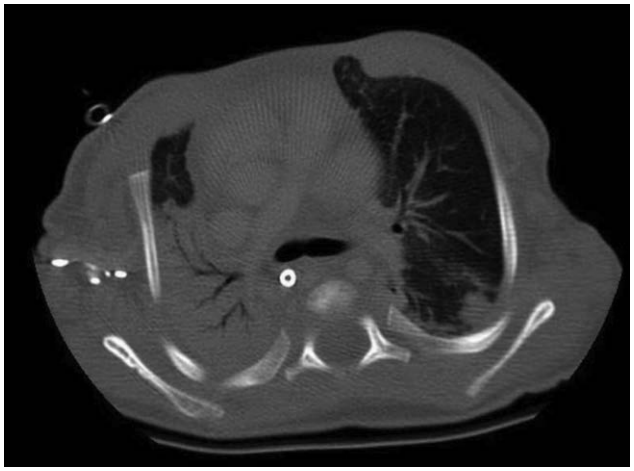

Fig. 3. Axial section of preoperative CT scan. Shows partial superior sternal cleft with distance between the manubrium sides of 4 cm together with partial herniation of the left lung and upper mediastinal structures through the sternal cleft.

and supraumbilical raphe (PHACES) syndrome,[15] Cardiac anomalies including Cantrell's pentalogy, among others.[16] Sternal clefts can also be an isolated entity, in which patients are usually asymptomatic, except when crying or coughing, which causes paradoxical chest wall movement (bulge during expiration and depression during inspiration).[2,12] Without surgical intervention, patients are at high risk of impaired oxygenation, frequent dyspnea and cough, as well as multiple chest infections.[3] This was evident in 1 of our patients, in whom paradoxical motion of the chest wall was associated with difficult weaning from ventilator support till further intervention to support the chest wall was applied.

Early surgical reconstruction of sternal cleft was recommended preferably in the neonatal period in which chest wall elasticity allows for primary approximation.[9–11] More complex interventions were recommended if such elasticity is lost or the characteristics of the cleft preclude primary cleft simple approximation like clavicle dislocation, chondrotomies, or the use of various autologous graft

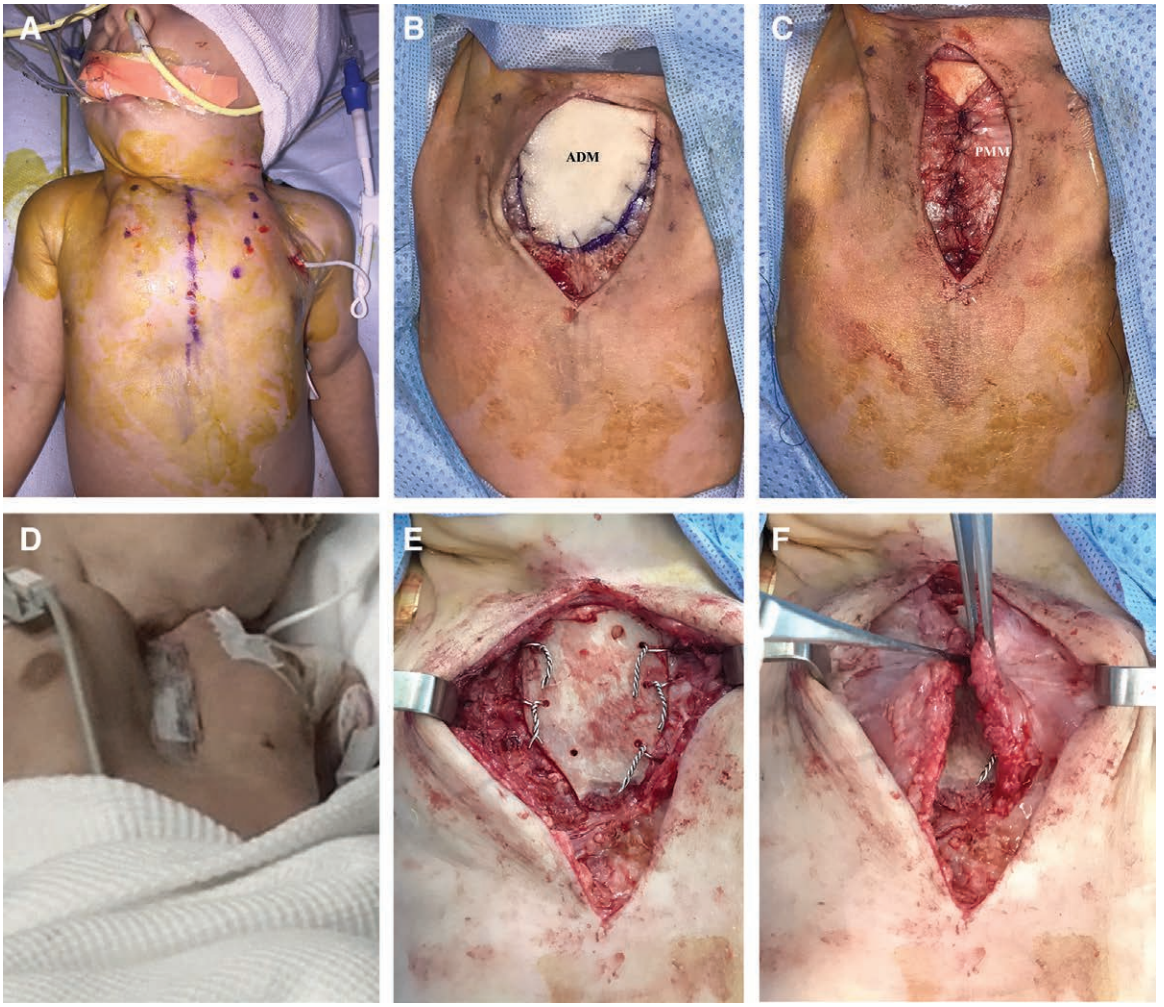

Fig. 4. Operative planning and reconstruction. A vertical incision line was made, followed by raising cutaneous and bilateral pectoralis major muscle flaps (A). SurgiMend dermal matrix graft was cut and secured to the cleft margins followed by approximation of bilateral pectoralis major muscle flaps (B and C). Postoperative complication of inward chest retraction is shown (D). Definitive skeletal reconstruction with allogeneic bone graft followed by bilateral pectoralis major advancement flaps (E and F). PMM, pectoralis major muscle; BG, bone graft.

options.[3,11,12] Mathevan et al.[17] suggested a treatment logarithm approaching patients with such deformities. Once a patient presented with sternal cleft, initial screening of certain associated anomalies depending on the location of cleft is warranted, for example, Cantrell's pentalogy to be associated with inferior type while the superior type (V or U shaped) to be associated with PHACES syndrome or various hamartomas. Radiological investigation involves CT scan with 3D reconstruction and magnetic resonance imaging of the chest. Other modalities like abdominal ultrasonography and echocardiography to check associated anomalies were recommended. Once a patient was optimized for surgical reconstruction, primary approximation (before 3 months of age) was the preferred modality. An intraoperative trial of approximation with no associated cardiovascular compromise was recommended before definitive primary approximation and closure. Beyond that age frame or if patients were in need for additional surgical procedures (cardiac or diaphragmatic defect repair), complex sternal defect reconstruction is warranted.[11,17]

In our patients, primary closure was not feasible, and hence other reconstructive options were considered. Definitive skeletal reconstruction was achieved with the use of an autogenous rib graft in 1 patient, and allogeneic bone graft in the other patient with no complications. The use of ADM substitutes in sternal cleft reconstruction was tried in the literature with promising outcomes. In one report, a superior V-shaped sternal cleft with 34 mm diastasis of the manubrium in a 4-year-old baby girl was reconstructed with XCM Biologic tissue matrix (Ethicon, Inc.) followed by bilateral pectoralis major muscle advancement flaps. The patient tolerated the procedure and was discharged on the eighth day postoperatively with no complications or abnormal chest wall excursion described.[13] Others have reported staged sternal reconstruction in 2 patients, initially with GorTex mesh (W.L. Gore & Associates, Inc.) and resorbable plate (Synthes Inc.) followed by pectoralis major muscle flap at 4 and 8 months of age. This was followed by definitive reconstruction at 3 years of age with the removal of GorTex mesh, resorbable plate, and application of Synthes ultrathick DermaMatrix (Synthes Inc.). Both patients tolerated the procedure well, with seroma formation during subsequent follow-ups.[14] We have used a similar approach after an intraoperative trial that showed difficulty in harvesting an autologous rib graft affecting chest wall stability as discussed with the interdisciplinary team. Dermal matrix graft of higher mechanical properties (SurgiMend, Integra LifeSciences Inc.) was then utilized as a means to stabilize the cleft margin in 1 of the patients. This was unfortunately associated with lack of a stable chest wall excursion, with return of abnormal paradoxical motion creating higher oxygen requirement for the patient. Definitive stable reconstruction was achieved later on with the use of allogeneic bone graft with no associated complications.

The use of ADM in the reconstruction of sternal clefts should be approached with caution considering the location of the cleft as well as the associated chest wall dynamics. Such use can aim in the soft-tissue reconstruction rather than skeletal stability.

Surgical reconstruction of sternal clefts represents a challenging situation for both the operating surgeon and the monitoring anesthetist. Rapid blood loss, arrhythmias, cardiac dysfunction, and pneumothorax are all possible complications associated with the surgical repair of sternal clefts. Advanced monitoring including transesophageal echocardiographic probe and other means was recommended to measure cardiovascular function throughout the procedure.[18] Planning with other reconstructive team members was of great value, especially before making incisions in which infiltration with diluted epinephrine solution was necessary to minimize associated blood loss, then again during rib graft harvest and allogeneic bone grafting.

CONCLUSION AND SUMMARY

Sternal clefts are rare congenital chest wall anomalies with variable presentations. Meticulous patient assessment and screening for any associated anomalies are crucial in determining the overall prognosis. Treatment also requires a multidisciplinary approach. Early surgical intervention to repair the cleft is recommended in the neonatal period of the child's life (preferably before 3 months of age), allowing for primary cleft closure. Surgeon's communication with a specialized pediatric anesthetist helps to plan and improve the overall surgical outcome. The use of different ADM products in the reconstruction of sternal clefts represents an interesting modality to aid and augment the surgical repair applied, but should be approached with caution.

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