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Discovery and Management of Diaphragmatic Hernia Related to Abandoned Epicardial Pacemaker Wires in a Pregnant Woman with {S,L,L} Transposition of the Great Arteries

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Abstract

Epicardial pacemaker leads placed during childhood are often not removed when transvenous systems are placed later in life. The risk of complications related to retained pacemaker leads and generators is not clear but is generally considered low. We report the case of a 23-year-old pregnant woman who presented with left upper quadrant pain at 20 weeks gestation. The patient was born with {S,L,L} transposition of the great arteries and had high-grade conduction disease in infancy compelling epicardial pacemaker placement. A standard transvenous pacemaker was placed at age 9 years, without removal of the epicardial system. The patient's abdominal pain was attributed to herniation of abdominal contents through a diaphragmatic defect at the site of the abandoned epicardial pacing wire. Her pain improved spontaneously but worsened later in pregnancy leading to repair of the diaphragmatic hernia via anterolateral thoracotomy at 30 weeks gestation. The procedure was well tolerated by mother and fetus. At 38 3/7 weeks gestation, the patient underwent uneventful delivery by cesarean section for breech presentation. This case illustrates the importance of multidisciplinary collaboration in the care of women with congenital heart disease.

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Keywords

Pacemaker; Transposition of Great Vessels; Diaphragmatic Hernia; Pregnancy

Introduction

{S,L,L} transposition of the great arteries, also referred to as congenitally or physiologically corrected or L-loop transposition of the great arteries, is characterized by atrioventricular and ventriculoarterial discordance.¹ {S,L,L} TGA is associated with a markedly increased incidence of complete atrioventricular (AV) block.² A reported 27% to 45% of patients with {S,L,L} TGA require pacemaker placement.³ Due to size and growth constraints, pacemakers inserted during infancy often use surgically placed epicardial leads with wires that traverse the diaphragm to connect to an abdominal generator.⁴ We report a case of diaphragmatic hernia at the site of abandoned epicardial pacer wire in a pregnant woman with {S,L,L} TGA.

Case Presentation

A 23-year-old woman with {S,L,L} TGA was referred to an adult congenital heart clinic for evaluation and management in the setting of an intrauterine pregnancy (G₁P₀). She had required no intracardiac surgery and had been generally asymptomatic. She was known to have a dysplastic left-sided tricuspid valve with moderate tricuspid regurgitation. Her only prior procedures related to cardiac conduction disease. The patient was noted to have had variable AV block that developed at 6 months of age leading to placement of an epicardial pacemaker system. A right ventricular epicardial lead was placed via left inframammary crease anterolateral thoracotomy with the wire traversing the diaphragm to the generator located in the left upper quadrant of the abdomen. At age 9 years, generator charge depletion of the abdominal pacemaker prompted the placement of a transvenous dual chamber pacemaker via a left subclavian vein with the generator placed subcutaneously in the left chest. The epicardial pacemaker wire and generator were not removed, and medical records from that time are not available to investigate the reasoning behind these decisions.

The patient first presented to our clinic at 15 weeks gestation. She had no abdominal pain or symptoms suggestive of heart failure or arrhythmia. Physical examination was relatively unremarkable with a right brachial blood pressure of 110/69 mm Hg, regular heart rate of 76 beats per minute and an oxygen saturation of 99% without supplemental oxygen. Lungs were clear to auscultation. The transvenous pacemaker generator was positioned normally in the left upper chest without erythema or tenderness. She had a normal S1 and a single S2 with a grade 2/6 holosystolic murmur at the left lower sternal border radiating to the apex. Her abdomen was soft and not tender to palpation. She had no edema.

The electrocardiogram revealed sinus rhythm at 79 beats per minute with marked first-degree AV delay (PR duration 450 ms). Pacemaker interrogation revealed that only 6% ventricular paced beats (the transvenous atrial lead had previously fractured, and the device was set to VVIR mode with a backup rate of 60). Bicycle cardiopulmonary exercise testing demonstrated normal heart rate and blood pressure response to exercise. Peak VO₂ was 18.5 mL/kg/min (55% predicted) with peak respiratory equivalent ratio of 1.24 and a normal Ve/VCO₂ slope. A transthoracic echocardiogram confirmed a tethered, dysplastic tricuspid valve with moderate regurgitation and mildly impaired systemic right ventricular function. Left ventricular function was normal without evidence of pulmonary hypertension. There was no ventricular septal defect or left ventricular outflow tract obstruction.

At 20 weeks gestational age, the patient presented with severe cramping left upper quadrant pain. She underwent a pelvic ultrasound showing avascular heterogeneous globular regions on the anterior surface of the placenta measuring $43 \times 30 \times 28$ mm and $29 \times 28 \times 26$ mm raising a question of placental abruption. These findings were new since the ultrasound performed 1 week prior. However, the left upper quadrant location of the pain argued against this being the direct cause of the patient's symptoms. Given her severe unremitting pain, computed tomography (CT) of the abdomen and chest was performed revealing a diaphragmatic hernia located where her epicardial pacer wires crossed the diaphragm (Figure 1). The appearance was suggestive of an iatrogenic diaphragmatic hernia, and less consistent with other types of diaphragmatic hernia (e.g., Morgagni's, Bochdalek, or hiatal). There was no evidence of bowel obstruction or ischemia, and conservative management with bowel rest and close observation for possible bowel obstruction or ischemia was instituted. Her symptoms spontaneously resolved following the CT scan. Consideration was given to immediate repair of the diaphragmatic hernia, but this was deferred due to her clinical stability. She was discharged on hospital day 4 with a plan for elective postpartum surgical repair.

The patient, at approximately 30-weeks gestational age, presented with left upper quadrant abdominal pain, now associated with nausea and vomiting. Vital signs were normal but examination was notable for a distended but soft abdomen with severe left upper quadrant tenderness. Jugular venous pressure was normal, and there was no edema or other examination evidence of heart failure. Laboratory evaluation was unremarkable, with a normal white blood cell count, creatinine concentration, and lactate and bicarbonate concentrations. She was again initially managed conservatively with bowel rest and pain medication. However, the pain persisted and on hospital day 5, a repeat abdominal CT demonstrated a larger diaphragmatic hernia with a 20 cm segment of colon in the left chest (Figure 2). The pain became progressively more severe, and the patient was started on total parenteral nutrition on hospital day 10. At this time, the decision was made to perform diaphragmatic hernia repair.

On the 12th hospital day, she underwent a left anterolateral thoracotomy repair of the diaphragmatic hernia under general endotracheal anesthesia. Preoperatively, a T8-T9 thoracic epidural and a right radial arterial line were placed. In the operating room, she was positioned supine with left uterine displacement. Transabdominal fetal heart rate monitoring was established in the preoperative area and continued throughout the case and into immediate postoperative period.

After preoxygenation, rapid sequence induction and intubation was performed. An 8.5 single lumen tube was placed without difficulty, and then exchanged to a 37 left double lumen tube. The patient was positioned in right lateral decubitus and one-lung ventilation was instituted with serial arterial blood gas monitoring. An exploratory thoracoscopy was performed with ports in the sixth intercostal space at the tip of the scapula and eighth intercostal space in the posterior axillary line. The hernia sac was noted to be pedunculated posteriorly with the actual neck positioned anteriorly. An anterolateral thoracotomy was performed. The diaphragm was incised just anterior to the hernia neck, and the hernia sac was circumferentially freed around the diaphragmatic defect. The pacer leads were noted to wrap around the colon and diaphragmatic defect. The leads were divided and capped and dissected free of the defect. The diaphragmatic defect had to be enlarged to allow reduction of the hernia into the abdomen. There was no evidence of bowel perforation, ischemia or injury, and the diaphragmatic opening was closed with a combination of primary suture repair and two-ply mesh. General anesthesia was maintained with sevoflurane, fentanyl, and vecuronium, which were chosen based on their long record of use during pregnancy. At the end of the case, double-lung ventilation was reestablished, muscle paralysis reversed,

ondansetron was given for nausea prophylaxis, the epidural was bolused with bupivacaine, the stomach and oropharynx were suctioned, and the patient was extubated when awake and following commands. There were no fetal heart rate decelerations or uterine contractions. The patient and fetus tolerated the operation well without complications. Fetal ultrasound postoperatively was reassuring. The patient was discharged on postoperative day 6.

The remainder of her pregnancy was uneventful. She presented with rupture of membranes at approximately 38 3/7 weeks gestation and underwent Cesarean section due to fetal breech presentation. Apgar scores were 7 at 1 minute and 9 at 5 minutes.

Discussion

We describe the case of a woman with {S,L,L} TGA who presented during pregnancy with a symptomatic diaphragmatic hernia related to abandoned epicardial pacemaker leads. Several studies have explored the long-term functional outcome of epicardial leads in children,⁵⁻⁹ but little has been reported about potential complications due to abandoned epicardial leads placed during childhood. In adults, abandoned *transvenous* pacemaker leads appear to carry a low though finite risk for complications, most commonly venous thrombosis and infection. A small study of 60 adults with an abandoned, noninfected epicardial leads found 20% were associated with a complication (lead migration 5, skin erosion 3, venous thrombosis 2, muscle stimulation 2).¹⁰ Another reported rare complication is cardiac strangulation by abandoned epicardial leads.¹¹ Intrapericardial diaphragmatic hernia is uncommon and usually related to prior trauma or iatrogenic causes.¹² Two cases of intrapericardial diaphragmatic hernia related to epicardial pacing leads have been reported.¹³

In the reported case, the increased intra-abdominal pressure during pregnancy likely precipitated or exacerbated herniation of abdominal contents through the transdiaphragmatic course of the abandoned epicardial wire. The potential for pregnancy to affect present pacemaker leads in other ways is highlighted by a recent case report of complete transection of epicardial pacing wires during pregnancy.¹⁴ Historically, in infants and in children, epicardial pacemaker leads were usually placed via thoracotomy, and the abdominal generator was placed via an abdominal incision. The surgical approach has evolved, and placement of epicardial leads and generators are now commonly performed via a subxiphoid or subcostal approach, thereby avoiding leads traversing the diaphragm.¹⁵ These approaches may eliminate the risk of diaphragmatic hernia described in this report.

Diaphragmatic hernia during pregnancy, while rare, may be catastrophic and associated with high mortality.^{16,17} A recent report and review described a total of 39 published cases since the first reported case in 1913 of a fatal outcome of a strangulated diaphragmatic hernia during pregnancy and puerperium.¹⁶ Just over half of these cases presented during pregnancy with the others presenting during delivery or postpartum. Of these 39 reported cases, 10 resulted in maternal death. Death was often associated with strangulation of herniated organs or mediastinal shift, and was more common among women presenting in labor or immediately postpartum, and 7 of the 10 maternal death cases were managed non-operatively. It is hypothesized that increased uterine size and, during active labor, contraction of the diaphragm precipitates or aggravates abdominal viscera strangulation, diaphragmatic rupture, or cardiopulmonary collapse.¹⁶⁻¹⁸ Due to the high reported mortality rate, some suggest that prophylactic operative repair of the defect should be considered for all pregnant women who present before the third trimester, even for those who are asymptomatic. In general, surgical repair is indicated for symptomatic patients.^{16,19} In cases that present in the third trimester or when delivery is imminent, cesarean section simultaneously with diaphragmatic repair may be the preferred management approach.^{16,19} The reported cases undoubtedly represent a biased sample, however. Diaphragmatic hernia

(exclusive of hiatal hernia) may be more common than appreciated, and the true risk of complications and death is unclear. Indeed, most of the women who had complications of diaphragmatic hernia during pregnancy had prior uncomplicated pregnancies.¹⁶

Pregnancy is generally well tolerated among women with {S,L,L} TGA, but there is an 11% to 13% reported complication rate.^{20,21} Complications typically relate to increased intravascular volume and cardiac output during pregnancy, leading to higher demands on the systemic right ventricle and exacerbation of systemic tricuspid regurgitation. In our case, these considerations as well the patient's pregnancy with a viable fetus complicated surgical repair. There was considerable discussion about the best approach to the patient's care among several hospital consultation services including maternal fetal medicine, general surgery, thoracic and cardiac surgery, anesthesia, electrophysiology, and adult congenital heart disease. While there are case reports of general anesthetic management of patients with {S,L,L} TGA,²² and of pregnant patients requiring thoracotomy and thorascopic procedures,²³ this, to our knowledge, is the first report to describe a case of a pregnant woman with {S,L,L} TGA who underwent successful thoracotomy.

Preoperative anesthetic concerns included the relatively modest risk directly related to the patient's congenital heart disease in the setting of preserved systemic ventricular function and no more than moderate atrioventricular valve regurgitation, as well as the impact of normal cardiovascular physiologic changes of pregnancy (increase in blood volume and cardiac output, decrease in systemic vascular resistance, aortocaval compression) and surgical requirements (lateral decubitus position, with lung isolation/one-lung ventilation, and profound muscle relaxation for diaphragmatic dissection) in the setting of a viable fetus. Additional considerations were increased risks for aspiration, difficult endotracheal intubation, or hypoxic episodes associated with intubation related to the presence of intrathoracic incarcerated bowel, in addition to the decrease in esophageal sphincter tone, mucosal engorgement/friability, decreased functional residual capacity, and increased oxygen consumption associated with pregnancy. General anesthetic principles for non-obstetric surgery in the obstetric patient, including prevention and aggressive treatment of hypotension, hypoxia, acidosis or alkalosis, and aortocaval compression were followed.²⁴

In summary, this case describes the rare complication of diaphragmatic hernia at abandoned epicardial lead site and its successful multidisciplinary management in a pregnant woman with {S,L,L} TGA. Close collaboration between adult congenital heart disease, maternal fetal medicine, general surgery, thoracic surgery, and anesthesiology teams facilitated an excellent outcome. Nevertheless, as an increasing number of women with complex congenital heart disease are reaching childbearing age, complications due to abandoned transdiaphragmatic abdominal epicardial wires may become a more common issue.

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Figure 1. Scout film from CT of chest and abdomen showing location of abdominal pacemaker generator (black arrow) and abandoned epicardial leads (white arrows). The asterisk identifies herniated bowel.

Figure 2.

(A) Frontal projection of lower thorax and upper abdomen showing herniation of bowel (asterisk) into thoracic cavity through diaphragmatic defect (white arrow). The bright signal on the right border of the defect represents retained pacemaker lead. (B) Axial projection showing bowel loops (short white arrows) within the defect bounded medially by pacemaker lead.