



Concomitant transsternal repair of a congenital Morgagni hernia and ventricular septal defect in a patient with Down syndrome

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ABSTRACT

Congenital Morgagni hernias are uncommon diaphragmatic hernias. Some cases of congenital Morgagni hernias are associated with congenital malformations, including congenital heart defects, chest wall abnormalities, and some chromosomal anomalies. Congenital Morgagni hernias should be treated surgically via transthoracic or transabdominal approach. However, in patients undergoing open heart surgery for congenital heart defects, transsternal exposure and repair can be also used. Herein, we report a case of a ventricular septal defect and congenital Morgagni hernias associated with Down syndrome of which the defects were repaired concomitantly via sternotomy approach.

Keywords: Morgagni hernia; transsternal repair; ventricular septal defect.

Congenital Morgagni hernias (CMH) are uncommon diaphragmatic hernias comprising about 3 to 5% of all types of congenital diaphragmatic hernias.^[1,2] Some cases of CMH are associated with congenital malformations, including congenital heart defects (i.e., atrial and/or ventricular septal defects, patent ductus arteriosus), chest wall abnormalities, and some chromosomal anomalies Down syndrome being the most common.^[3-5] Once diagnosed, surgical repair is indicated by either transabdominal or transthoracic approach. We, herein, report a case of a ventricular septal defect (VSD) and CMH associated with Down syndrome of which the defects were repaired concomitantly via sternotomy approach.

CASE REPORT

An 11-month-old male with Down syndrome was referred to our institution upon detection of cardiac murmur on physical examination. On chest X-ray, gas-filled loops of the bowel were located within the right chest cavity. Lateral chest X-ray confirmed the diagnosis, showing a retrosternal radiolucent shadow. Echocardiography revealed a 4.5 mm subaortic ventricular septal defect with slight malalignment and a small secundum atrial septal defect. Chest computed tomography scan identified a 1.5×2.7 cm diaphragmatic defect in the

right cardiophrenic sinus and loops of bowel within the chest and confirmed the diagnosis of CMH (Figure 1).

We decided to close VSD and to repair the CMH concomitantly. Midline sternotomy was performed. Cardiopulmonary bypass (CPB) was instituted in a routine manner by aortic and bicaval cannulation. The VSD was closed using a Dacron® patch via right atrium. The patient weaned from CPB. Following completion of the cardiac procedure, right parietal pleura was opened. The diaphragmatic defect and the hernia sac which consist the transverse colon inside were visualized anteriorly close to the pericardiac fat. The hernia sac was dissected and lowered to the abdomen throughout the defect. The defect was repaired with a Dacron patch with interrupted sutures. Postoperative period was uneventful. The patient was discharged on postoperative Day 10.

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Figure 1. A 1.5×2.7 cm diaphragmatic defect in the right cardiophrenic sinus and loops of bowel within the chest.

DISCUSSION

Anterior congenital diaphragmatic hernia through the foramen of Morgagni was first described by Giovanni Morgagni in 1761.^[1,6] The defect results from the failure of the fibrotendinous portion of the pars sternalis to fuse with the fibrotendinous part of the costochondral arches.^[1,2,6] Most (%90) CMH occur on the right side, 2% on the left side, and 8% occur bilaterally. Pericardial attachment to the diaphragm, which is more common on the left side, supports and protects that side. The most common contents of the hernia sac are the colon, small bowels, liver, omentum, and stomach.^[6]

It is well-known that CMH may be associated with other congenital anomalies such as congenital heart diseases, Down syndrome, intestinal malrotation, omphalocele, pectus carinatum, and genitourinary anomalies.^[3] However, chromosomal anomalies which are associated with CMH are more significant, in terms of whether it is an inheritable defect.^[7,8] In large series, it was reported that the incidence of CMH associated with Down syndrome was approximately 20%.^[3] It has also been reported that there is a possibility of muscular deficiency of the ventral paramedian segment of the body wall related to the diaphragm in Down syndrome.^[9]

There is a common consensus which CMH should be treated surgically in symptomatic patients. Opinions differ as to the best surgical approach, whether

transthoracic or transabdominal.^[10,11] However, in patients undergoing open heart surgery for congenital heart defects, as in our case, transsternal exposure and repair is also a possible option.^[12] Another controversial issue is the removal of the hernia sac during repair. Although some authors recommend excision of the hernia sac, others prefer to leave it as to avoid the possibility of pneumopericardium.^[3,13] However, there is no available data in the literature indicating whether leaving a hernia sac influences the rate of recurrence.

In conclusion, as congenital Morgagni hernias can be associated with congenital cardiac diseases, it should be kept in mind, in patients undergoing open heart surgery via sternotomy approach, that these defects can be repaired with transsternal approach as effective as other approaches without an additional incision.

Declaration of conflicting interests

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