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Case report

Concomitant trans-sternal repair of Morgagni hernia and ventricular septal defect in a patient with Down syndrome: A case report

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ABSTRACT

Introduction: Morgagni hernia is a rare type of hernia occurring secondary to potential anterior-medial defects in the diaphragm. The association of the defect with congenital cardiac pathologies and Down syndrome are well known. The defect is repaired usually by trans-abdominal or transthoracic approaches. Trans-sternal repair of the hernia is preferred in patients undergoing concomitant open heart surgery.

Case presentation: A 2-year-old child with Down syndrome underwent concomitant repair of Morgagni hernia and closure of his ventricular septal defect under cardiopulmonary bypass. The hernia was corrected by the sternotomy approach, without opening the hernia content, before the correction of the cardiac pathology. The patient made an uneventful recovery and was discharged on the 4th postoperative day.

Discussion: Preoperative diagnosis of diaphragmatic hernia in congenital heart disease is important to decrease mortality rate. However, trans-sternal exposure of the defect is also possible, as in this case, in patients undergoing open heart surgery for congenital cardiac defects. The defect can be repaired by this approach, concomitantly with the cardiac anomaly, no need for an additional incision and without opening the hernia sac.

Conclusion: Our experience, although very limited, in patients who are suffering from Morgagni hernia and concomitant congenital heart defects shows that simultaneous repair of Morgagni hernia through midline sternotomy prior to cardiac procedure is effective. As Morgagni hernia can be accompanied with many congenital cardiac anomalies, cardiac surgeons should be familiar with the trans-sternal approach to the defect.

1. Introduction

Morgagni hernia is a rare type of hernia occurring secondary to potential anterior-medial defects in the diaphragm [1]. This anomaly usually diagnosed incidentally on a chest radiograph ordered for some other reasons, commonly a respiratory complaint. Some cases of Morgagni hernia can be associated with congenital heart defects (atrial and/or ventricular septal defects, patent ductus arteriosus), chest wall abnormalities, and some genetic syndromes, especially with Down syndrome [2–6]. The defect is usually repaired by a transabdominal or transthoracic approach. Based on the **S**urgical **Ca**se **Re**port, 2020 (SCARE) guidelines, here, we report the concomitant repair of Morgagni hernia by a sternotomy approach in a child with Down syndrome who had undergone open-heart surgery for the closure of a ventricular septal defect [7].

2. Case presentation

A 2-year-old male child with Down syndrome was referred to Mellat Medical for Ventricular Septal Defect (VSD) closure by Afghan Red Crescent Society. While he had a positive family history of Down syndrome in his older brother, the social history and psychosocial history were unremarkable. While completing the routine preoperative investigations for VSD closure, suspicion of diaphragmatic hernia was

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noticed with the observation of the chest roentgenography of a possible herniation of the bowel loops into the right hemithorax (Fig. 1). An anterior diaphragmatic (Morgagni) hernia was established by CT-scan of the chest which contained both small and large bowel loops (Fig. 2). The two-dimensional color-flow Doppler echocardiography of the patient demonstrated a perimembranous ventricular septal defect (VSD) with inlet widening, pulmonary hypertension, and mild pericardial effusion. The decision for VSD closure and the concomitant repair of the diaphragmatic defect was then taken after discussion with the cardiac surgeon and pediatric general surgeon. During the operation, the thorax was opened by a vertical midline sternotomy. The peritoneal sac and the defect were seen after the parietal pleura was removed from the area to the right side. The hernia sac was incised from the pericardium and retrosternal space and was reduced to the abdominal cavity through the defect with the transverse colon in it (Fig. 3). Nylon, horizontal sutures were sequentially used through the edge of the diaphragmatic defect and into the retrosternal fascia and periosteum. There was no need to use mesh as the defect was not large and skin could be closed without tension as the sutures were tied. After the diaphragmatic hernia was repaired, the pericardium was opened and serous pericardial fluid was aspirated. Cardiopulmonary bypass was established in a routine manner by aortic and bicaval cannulation. VSD was closed from the right atrium approach with an autologous pericardial patch. The patient was taken from cardiopulmonary bypass upon completion of the cardiac procedure. The nasogastric tube was kept in place until the gastrointestinal (GI) function returned to normal. The postoperative postero-anterior and lateral roentgenograms of the patient showed the disappearance of the mediastinal shadows (Fig. 4). After 4 days of hospitalization, the patient made an uneventful recovery and was discharged with normal GI function. At 3 months follow-up the patient is healthy, vital signs are within normal limits and echocardiography results are satisfactory VSD closure with normal biventricular function.

3. Discussion

Morgagni hernia is a rare type of diaphragmatic hernia in the pediatric age group and is often uncommon with congenital diaphragmatic hernia (CHD) (VSD). Its frequency varies from 1% to 9.6% in large studies [2,8]. The foramen of Morgagni is located in the retrosternal space which results from the failure of the fusion of the fibrotendinous part of the pars tendinalis arising from the costochondral arches with the fibrotendinous portion of the par sternalis. This space is usually filled with fat and covered by pleura from the top and peritoneum inferiorly. When present, the defect shows a path through which abdominal viscera can herniate into the chest. Morgagni hernia usually takes place at the anteromedial part of the junction of the thoracic wall and septum transversum. In most of the cases (90%) the defect is right-sided while the remaining defects are left-sided or bilateral [9]. Morgagni hernia is usually asymptomatic and can be seen that there is no bowel in the defect, otherwise the risk of incarceration is an urgent indication for operating room. The colon, small bowels, liver, omentum and stomach are the most contents of the defect. In a repeated manner, it causes respiratory discomforts due to the compression of the lower lobe of the ipsilateral lung. There are up to 26% of cardiac anomalies associated with CHD [10]. Of every 5 liveborn with Morgagni hernia, 3 had trisomy 21. The considerable association between the hernia and Down syndrome is possibly due to defective dorsoventral migration of rhabdomyoblasts from the paraxial myotomes, caused by an increased cellular adhesiveness in trisomy 21 [6]. The transabdominal route of repair is the most welcomed method but the transthoracic approach by a limited thoracotomy is advocated if the herniated sac has solid contents [2]. Recently, laparoscopic techniques have obtained significant attention and are considered as an alternative approach for diaphragmatic hernias [11]. However, trans-sternal exposure of the defect is also possible, as in this case, in patients undergoing open-heart surgery for congenital cardiac defects. The defect can be repaired by this approach, simultaneously with the cardiac anomaly. As Morgagni hernia can be accompanied by many congenital cardiac anomalies, cardiac surgeons should be familiar with the trans-sternal approach to the defect which is as effective as other surgical approaches.

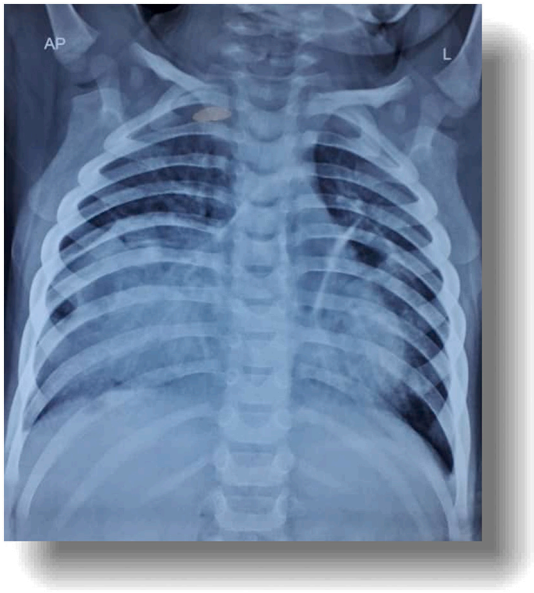

Fig. 1. The chest X-ray before the procedure.

4. Conclusion

Our experience, although very limited, with patients suffering from Morgagni hernia and concomitant congenital heart defects reveal that simultaneous repair of Morgagni hernia through midline sternotomy prior to the cardiac procedure is safe and effective.

Consent

Informed consent was obtained from the patient`s legal guardian (his mother), for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

This is a case report paper.

Funding

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Guarantor

Sayed Hamid Mousavi, the corresponding author, accepted full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Research registration number

Not applicable.

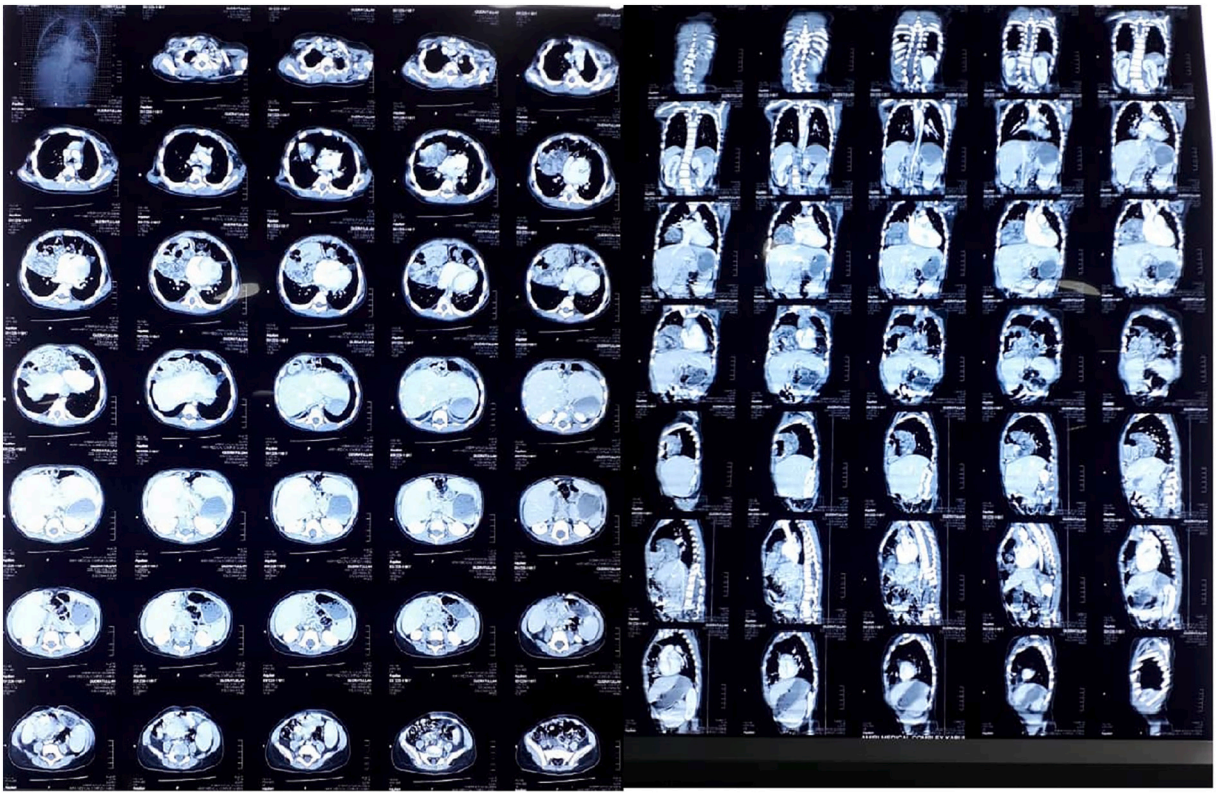

Fig. 2. These Images show a large anterior diaphragmatic (Morgagni) hernia with contained small and large bowel loops.

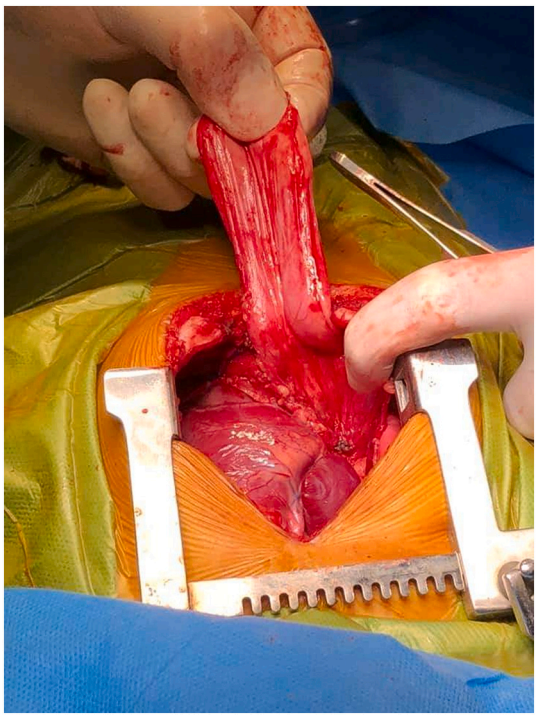

Fig. 3. The photo shows that hernia sac was dissected from the pericardium and retrosternal space and was reduced to the abdominal cavity through the defect with the transverse colon in it.

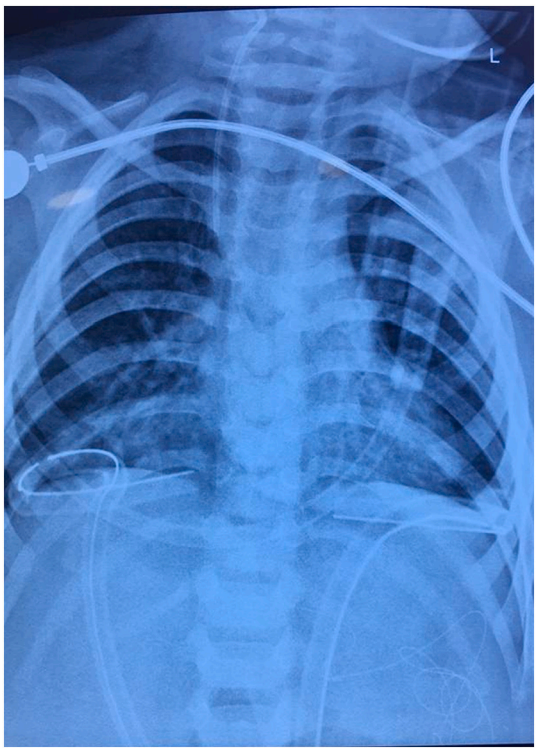

Fig. 4. This is a postoperative X-ray which shows the disappearance of the mediastinal shadows.

CRediT authorship contribution statement

AAD, HGT and MAR conceived and designed the study; AFW and HGT wrote the manuscript; MAR and FN helped collect data;; SHM confirmed the eligibility of the participants' for the study; SHM and FN Supervised the whole study and approved the final version of the

manuscript.

Declaration of competing interest

The authors report no declarations of interest.

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