Successful surgical management of combined presence of congenital diaphragmatic hernia, ventricular septal defect, and patent ductus arteriosus in a child with allergy to frusemide—a case report

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Abstract

A 6-week-old male child presented with a right-sided congenital diaphragmatic hernia (CDH), large perimembranous (PM) ventricular septal defect (VSD) and multiple muscular VSDs, and a patent ductus arteriosus (PDA) with severe pulmonary arterial hypertension (PAH). The child was found to be allergic to frusemide. The VSDs were closed, the PDA was ligated, and the CDH was repaired through a right subcostal incision. Postoperatively, there were recurrent episodes of right lung lobar collapse, but the child responded to intravenous antibiotics and nasal continuous positive airway pressure (CPAP). The child is medically stable and on outpatient follow-up. We present this rare combination of diseases in view of a successful simultaneous surgical repair.

Keywords Frusemide allergy · Combination of VSD · Single-stage combined surgical repair

Introduction

CDH has an approximate incidence of 1 in 3000 live births [1]. Associated anomalies are present in 40–50% cases [1]. Among them, congenital heart anomalies are prognostically the most important. However, data on the outcome are limited on this association and confined mainly to case reports or series [2]. We are reporting a treated case of combined presence of CDH and VSD along with PDA with associated allergy to frusemide in view of limited documented literature of such cases and the success achieved in the management.

Case report

In December 2015, a 6-week-old male child presented to us with a recent history of regurgitation of feed associated with respiratory distress. On clinical evaluation and contrast-enhanced computed tomography (CECT) chest and abdomen, he was diagnosed with a right-sided CDH with collapsed middle and lower lobes of right lung (Figs. 1, 2 and 3). The child was admitted in intensive care unit (ICU) and had to be put on mechanical ventilation. The child was not on prostaglandin infusion. Initially, in the emergency until the point of shifting to the ICU, the child was on artificial manual breathing unit (AMBU)-assisted hand ventilation. In the initial phase, we could not arrange a bed with ventilator and the child had to be kept in emergency in suboptimal conditions. This and the high pulmonary artery (PA) pressures possibly led to haemodynamic compromise and also borderline renal function. Subsequently, in the ICU, pressure-regulated volume control ventilation was used. However, high-frequency oscillatory (HFO) ventilation was not used for this child. A 2D echocardiography was performed which showed 8–

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CDH was subsequently approached through a right subcostal incision, the herniated bowel and liver was retracted back into the abdomen, and the diaphragmatic defect was closed with interrupted 2–0 silk sutures. We used nonabsorbable sutures to repair the diaphragmatic defect and avoided polypropylene as for secured knotting polypropylene sutures would require multiple throws and the protruded sharp edges of the polypropylene could damage the underlying liver for the young child.

Postoperatively, the child was kept under elective mechanical ventilation for about 16 h and sildenafil was administered which further lowered the elevated pulmonary arterial (PA) pressure allowing subsequent extubation. Sildenafil was added to the postoperative medical management as even after initial lowering of the PA pressures immediately postoperatively, the child had intermittent episodes of high PA pressures as diagnosed by serial postoperative echocardiography in ICU. This also helped us to wean off the intravenous inodilator therapy in the form of milrinone and dobutamine. We had started milrinone initially and then dobutamine was started and milrinone was tapered off. This was done in view of the decreasing platelet counts for the patient which is a documented side effect of milrinone and keeping in mind the high cost of milrinone, too. The postoperative course was stormy with recurrent episodes of right lung lobar collapse complicated by superadded lower respiratory tract infection which prompted a prolonged ICU stay. As already mentioned, to make the life of the managing team difficult, the child was also allergic to frusemide and the drug could not be used as a part of the decongestive medications. However, the child responded to intravenous antibiotics and nasal continuous positive airway pressure (CPAP) and vigorous chest physiotherapy. The postoperative echocardiography prior to discharge from the hospital showed no residual VSDs, no flow across the PDA or the PFO which we had left open, normal PA pressures, and normal biventricular function. The child was discharged from the hospital in a medically stable condition and presently is on outpatient follow-up.

Discussion

In the neonatal period, CDH presents with respiratory distress caused by insufficient oxygenation. The symptoms of insufficient gas exchange are coupled with those of persistent PAH due to arteriolar constriction and closure of the pulmonary arterial bed which results in a pattern of persistent foetal circulation whereby the blood from the right ventricle gets shunted to the left heart causing ineffective gas exchange [3]. Right ventricular dysfunction may result because of persistent PAH and in such circumstances, maintenance of PDA is indicated [4]. PM VSD, cardiac outflow anomalies, and abnormal great vessels are associated with about one-third of CDH patients [3].

A combined approach of delayed surgery and gentle ventilation provides the best results in CDH. The preoperative

measures are targeted to haemodynamically stabilise patient in the form of acceptable oxygenation ($\text{PaO}_2 > 40$ mmHg) and CO_2 disposal ($\text{PaCO}_2 < 60$ mmHg) and stable pulmonary pressures ($< 50\%$ of systemic pressure), acceptable shunting, good myocardial function, and adequate renal clearance with minimal or withdrawn inotropic support. The treatment of PAH in these cases is of immense importance since it determines shunting of nonoxygenated blood out of the lungs.

In cardiac anomalies resulting in incomplete separation of the systemic and pulmonary circulation, such as the septal defects, there is shunting of blood from left to right which may lead to increased pulmonary flows and PAH. In a large VSD, the pulmonary arteries are exposed to systemic pressure immediately after birth and, hence, this causes greater degrees of early PAH which can be controlled by early repair [5].

However, medical stabilisation with decongestive medications including diuretic therapy helps to optimise patients with VSD prior to surgical therapy and also helps to defer surgery up to 6 months of age, which makes surgical closure less difficult.

Our patient had simultaneous presence of CDH as well as a large non-restrictive VSD with additional small VSD and a PDA which resulted in very high PA pressures with resultant difficulty in weaning off mechanical ventilation. Besides, the child being allergic to frusemide, medical optimisation with use of decongestive measures was not completely feasible. Hence we chose the path of combined surgical closure of VSD and PDA ligation along with repair of CDH as a single stage procedure to give the child more chances of survival with greater chances of reduction of postoperative PA pressures. The early approach of combined repair of VSDs and PDA ligation along with CDH resulted in lowering of PA pressures which either alone could not have provided.

Conclusion

We hope our case will add in the management of such cases in the days to come and we recommend combined early surgical repair of VSD, PDA, and CDH, especially in the associated unusual circumstance of frusemide allergy.

Compliance with ethical standards

Conflict of interest There is no conflict of interest among the authors.

Ethical approval All procedures performed in this case were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from the parents of the child included in the case report.

Written informed consent was also taken from the parents of the child for publication of the case report.

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