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CASE REPORT

Ventricular Septal Defect Repair in a Patient with Hydrocephalus

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ABSTRACT

Congenital heart diseases may be associated with different syndromes or anomalies. Congenital hydrocephalus is a rare disease that leads to increased intracranial pressure due to fluid accumulation through obstruction of intracranial aqueduct flow, Arnold Chiari and Dandy–Walker malformations, or abnormally increased fluid production into intracerebral ventricles. Increased intracranial pressure may produce many problems during the anaesthesia for any operation. Moreover, the postoperative period may be complicated by hydrocephalus. In severe cases, shunt replacement may be necessary. The case reported here is a 10-month-old infant with ventricular septal defect repair diagnosed with hydrocephalus and postoperative early follow-up findings.

Keywords: Ventricular septal defect, hydrocephalus, congenital heart disease

INTRODUCTION

The incidence of congenital hydrocephalus (CH) has been reported to be 0.5–0.8 per 1000 in the population (1). It results in undesirable fluid accumulation in the brain. The most frequent cause of CH is spina bifida, which is described as a multifactorial neural tube defect presented in a ratio of 3/100000 live births (2). In the case of spina bifida, meningomyelocele can be seen. Isolated congenital hydrocephalus may have a quiet normal outcome in general as high as 60 % (3). Poor outcome depends on the association with central nervous system anomalies in addition to the thickness of the cortical mantle (3). In the literature, there is limited acknowledgement that reports the challenging features of the CH associated with congenital heart disease (CHD) (3,4). The most related CHD's are patent ductus arteriosus, atrial septal defect, ventricular septal defect (VSD), coarctation of the aorta, tetralogy of Fallot, bilateral v. cava superior, bicuspid pulmonary valve, hypoplastic pulmonary valve, aortic atresia, dextrocardia, atrioventricular septal effect, and mitral and tricuspid insufficiency (3,4). Acyanotic CHD is frequent in this association (3). Management techniques in the operating room and postoperative period are challenging (3–5). In our case report, we present the successful repair of a VSD in a 10-month-old infant diagnosed with CH who had previously undergone surgical repair for meningomyelocele. During the VSD repair stage, CH was carefully monitored because of the undesirable clinical course worsening.

CASE REPORT

A 10-month-old infant was diagnosed with spina bifida and CH in a routine control. Further imaging tests were performed because of cardiac murmur. VSD was detected on the echocardiogram. In the first month, the patient underwent surgery for meningomyelocele. Postoperative follow-up was uneventful, and an echocardiogram was performed for the monitoring of VSD.

During follow-up, the patient had an insufficiency with swallowing reflex and was being fed via a nasogastric catheter. A ventriculoperitoneal shunt (VPS) was planned. However, the cardiac condition was found to be more of a priority than CH, so it was decided to perform VSD repair. Surgical indications were 10 mm perimembranous VSD, pulmonary artery hypertension of 40 mmHg, Qp/Qs >1,5 and frequent hospitalisation for recurrent respiratory tract infections. Informed consent was obtained from the patient's relatives.

Preoperative evaluation was performed, and no abnormality was detected in blood tests. The VSD was repaired using the transatrial

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approach. Induction of anaesthesia was made in normocarbia, normoxic, and normothermic situations, and vital parameters were within the normal range. Cardiopulmonary bypass (CPB) was established in mild hypothermia, and VSD repair was performed using a polytetrafluoroethylene graft. During CPB, brain circulation was monitored using near-infrared spectroscopy to maintain adequate blood supply. Postoperative blood loss was within an acceptable range, and thrombocyte suspension was not required. After 24 h, the patient tolerated the extubation, and the arterial tension was within the normal range. The neurological evaluation was normal. The possibility of intracranial haemorrhage was assessed by cranial ultrasound and was not found. The patient was discharged on the 10th day. No complications developed. Four months later, the patient underwent an operation for a VPS. Early follow-up for VSD repair was normal.

DISCUSSION

CH is frequently associated with central neurological system pathologies (6). The association between CH and Fallot tetralogy has been published in a case report (7). In addition, associated CHDs in CH have been reported in the literature as follows: VSD, atrioventricular septal defect, patent ductus arteriosus, coarctation of the aorta, atrial septal defect, congenitally corrected transposition of major arteries, and mitral and tricuspid valve regurgitations (3). Bicuspid aortic valve and pulmonary stenosis may be associated with CH (5). Double aortic arch has been reported as associated with CHD in only one case (8), and hypoplastic left heart and ectopia cordis were reported by Garne (1). Crawford stated in his study that CHD is strongly related to poor outcome in CH (3). Of the 11 children, 4 died early months in that series, and complex CHD added an additional risk for mortality even though corrective surgery was realised (3). Unfortunately, the mechanism of additional mortality risk was not mentioned in previous studies.

As Crawford stated, cardiac repair should be performed as a priority because of the cardiac condition of the patient (3).

Singh stated that left-to-right shunt due to double aortic arch, patent ductus arteriosus, and atrial septal defect might complicate the anaesthesia procedure for VPS operation (8). They preferred to manage the patient at the time of anaesthetic induction with hyperventilation to reduce cerebral blood flow while maintaining adequate PaO2 and avoided pulmonary vasodilation by lowering FiO2 and/or hypoventilation (8).

In some cases, surgical treatment of hydrocephalus requires a VPS operation; however, some case reports mentioned a requirement for a conversion of VPS to a ventriculoatrial shunt because of peritoneal fluid accumulation related to different factors (7). In these cases, an appropriate central venous pressure (CVP) ranged between 10 and 12 mmHg was obligatory to provide drainage of the excessive cerebral fluid (7). In case of right heart insufficiency and increased CVP, intracranial pressure could not be properly decreased. CHD by itself requires serious surgery and a long postoperative intensive care unit survey because of drainage after surgery, infections, pulmonary oedema caused by cardiopulmonary bypass (CPB) circulation. These postoperative reasons may trigger changes in acute phase reactant proteins, particularly serum albumin and protein levels. The cytokine storm after CPB generally leads to different degrees of capillary endothelium dysfunction. In conclusion, wide corporeal oedema and third space fluid accumulation due to extravasation occurred. In addition, as mentioned previously, increased CVP may lead to intracranial fluid accumulation (7). Therefore, in CH and CHD associations, intracranial fluid accumulation following cardiac surgery can be observed (9).

Moreover, CHD must be repaired under high-dose heparin for CPB. The risk of intracranial haemorrhage may be aggravated by thrombocyte dysfunction due to CPB circulation, thrombocytopenia, rebound phenomenon of heparin, perioperative arterial hypertension, or increased APTT and INR values due to acute liver dysfunction during postoperative monitoring. In our case, utilisation of high-dose heparin didn't result any intracranial haemorrhage and it is safe when careful perioperative follow-up is implemented. To the best of our knowledge, the association between CH and CHD is rare, and there are a few studies and limited case reports in the literature. It requires further studies to reveal the incidence of fluid accumulation after CHD surgery in patients with CH.

Informed Consent: Written consent was obtained from the participants.

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