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Anesthetic Management in an Infant with Dandy–Walker Syndrome Presenting with Acyanotic Heart Disease and Hydrocephalous Post-COVID-19 Recovery: A Rare Experience

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Abstract

We report a case of hydrocephalus with Dandy–Walker malformation in a 2-month-old girl child recently recovered from COVID-19. The child was detected to have acyanotic heart disease with left-to-right shunt and severe pulmonary arterial hypertension during the preoperative evaluation process for ventriculoperitoneal (VP) shunt placement. We share our experience of the perioperative management for pulmonary artery banding (PAB) and patent ductus arteriosus ligation as a part of staged cardiac corrective surgery, followed by VP shunt to relieve hydrocephalus in the single setting. Our management was focused on the preservation of the normal cerebral and cardiac physiology to prevent rise in intracranial pressure and pulmonary artery pressure. A multidisciplinary team, consisting of cardiac- and neuroanesthesiologists and cardiac and neurosurgeons, was involved in management of the case. Diligent maintenance of airway, stable hemodynamics, meticulous ventilation, along with postoperative ICU management helped in the successful outcome of this unique case.

Keywords
- Dandy–Walker syndrome
- acyanotic congenital heart disease
- hydrocephalus
- ventriculoperitoneal shunt
- anesthetic management

Introduction

Prevalence of Dandy–Walker malformation (DWM) is 5 to 8 per 100,000 live births.[1] In first year, 2 to 4% develop hydrocephalus and raised intracranial pressure (ICP).[1,2] Congenital heart disease is reported in 18%; association with tetralogy of Fallot is high.[3,4]

We share our experience of managing a case of hydrocephalus with Dandy–Walker disease (DWD) for ventriculoperitoneal (VP) shunt placement in a 2-month-old girl child who recently recovered from COVID-19. Although shunt surgeries are routine in pediatric patients, this case is unique because of preexisting, complicated cardiovascular anomalies, which had a direct bearing on the neurological outcome. Also, recent history of COVID-19 posed risk for unpredictable cardiopulmonary complications.

Case Report

A 2-month-old girl child, 5 kg, presented in neurosurgical department with history of gradually increasing head size, lethargy, poor feeding, and delayed milestone for 1 month. On examination, child was lethargic with classical features of hydrocephalus, sunsetting eyes, macrocephaly (50 cm), and

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bulging fontanelles. Noncontrast CT head revealed midline posterior fossa cystic lesion, cerebellar hypoplasia, severe hydrocephalus, with cortical thinning (►**Fig. 1**).

During preanesthetic evaluation for ventriculoperitoneal shunt surgery, her mother gave history of child's recent recovery from COVID-19 1 week back. Vitals (pulse rate—110/minute, blood pressure—110/69, respiratory rate—45/min, oxygen saturation on room air—96%) and blood investigations were normal. Airway was apparently normal, except mechanical issues with large head. Chest X-ray showed cardiomegaly; no radiological findings of COVID-19 pneumonia. Electrocardiogram showed normal sinus rhythm with P-pulmonale. 2D-echocardiography revealed double-outlet right ventricle (DORV), 6-mm subaortic ventricular septal defect (SAVSD), 3-mm patent ductus arteriosus (PDA), dilated pulmonary artery (PA), pulmonary arterial hypertension (PAH), and left-to-right-shunt. With these findings, a provisional diagnosis of Dandy–Walker variant with acyanotic heart disease with left-to-right-shunt and severe PAH was made. Pediatric cardiac surgery team reviewed the case and pulmonary artery banding (PAB), followed by VP shunt surgery was planned. High-risk consent was obtained from parents.

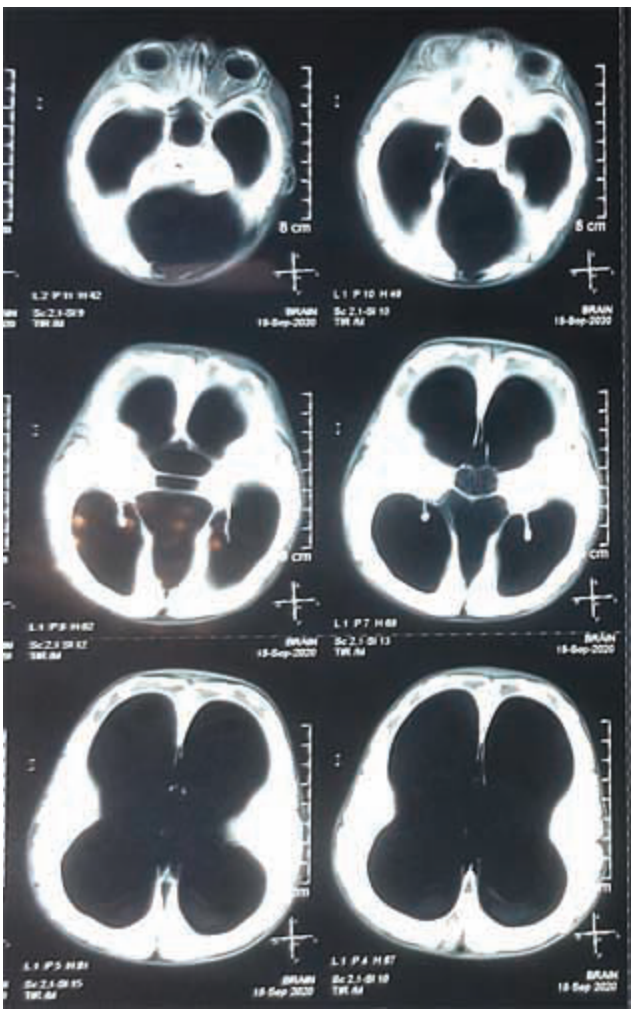

Fig. 1 CT scan showing posterior fossa cyst and hydrocephalus (large, dilated ventricles) with cerebral cortical thinning.

Maintenance fluid half normal saline and dextrose saline was started at 20 mL/hr with 6 hours fasting. Electrocardiogram leads, noninvasive blood pressure cuff, and saturation probe were applied, and baseline vital parameters were noted in the operation theater. Infant was premedicated with intravenous (IV) midazolam 0.5 mg and fentanyl 20 mcg on table under direct observation. Thiopentone 25 mg IV and a mixture of air, oxygen (1: 1) and sevoflurane (< 1 minimum alveolar concentration [MAC]) were used for anesthetic induction via closed circuit. A roll was placed below the child's shoulder, head on gelfoam ring, and body on an elevated platform (folded sheets), to facilitate endotracheal intubation after IV vecuronium 0.6 mg and 3-minute intermittent positive pressure ventilation (IPPV). Trachea was intubated with 3.5 mm ID flexometallic cuffed tube and fixed at 10 cm. Oropharyngeal temperature probe was used to monitor temperature. A 4.5F central venous access was obtained in the right femoral vein, and 22G arterial line in left femoral artery. Urobag was emptied; patient was handed over to cardiac surgical team. IV amikacin 30 mg and IV ceftriaxone 250 mg were administered before surgical incision. Infusion dobutamine at 5 mcg/hr and dopamine 5 mcg/ hr were started at thoracotomy and titrated to maintain mean blood pressure of 45 to 75 mmHg. Anesthesia was maintained throughout the surgery with air oxygen mixture (1:1), isoflurane 1.5 to 2%, and intermittent boluses of vecuronium and fentanyl.

PDA was ligated, and PAB was performed after thoracotomy. Transient fall in oxygen saturation (2–3%) from 98 to 99% with 50% FiO_2 followed PA tightening: sign of successful banding as volume of deoxygenated blood reaching lungs for oxygenation was reduced. A total of 100 mL of packed red blood cells (RBC) was transfused. Blood sugar was checked and recorded. After PAB, PA pressure was 26/14 mmHg with systemic pressure of 80/40 mmHg.

Postthoracotomy closure, position of the patient's head was turned 90 degrees to allow access for neurosurgery. Ventricular end of the shunt was inserted at the frontal horn of right lateral ventricle and free flow of cerebrospinal fluid (CSF) was obtained at the abdominal end.

Surgery lasted for 2.5 hours, total input was 80 mL, and urine output was 20 mL. Patient was shifted to cardiothoracic and vascular surgery (CTVS) ICU for overnight elective mechanical ventilation, sedated, and paralyzed. Next morning, infusion of vecuronium was stopped, infusion of fentanyl tapered off, and patient's trachea was extubated after blood gas analysis showed PaO_2 105 mmHg at FiO2 30%. Infusion of dopamine at 5mcg/kg/min and dobutamine 5mcg/kg/min was tapered and discontinued. The child remained in ICU for 3 days. Her sensorium improved, and she was discharged on postoperative day 10 from the ward active and accepting feeds.

Discussion

Anesthetic management was different from routine VP shunt surgeries because of the preexisting complex cardiac anomalies and a recent recovery from COVID-19, verified by negative reverse transcription polymerase chain reaction

(RT-PCR) report. Recovery from COVID-19 in children can be prolonged, and defining recovery, based on a negative RT-PCR report, can be tricky, given its low reliability in children. Pediatric COVID-19 disease can range from being asymptomatic with high viral load to multiorgan inflammatory syndrome. Perioperative concerns for surgery in such cases are false-negative RT-PCR in children, low yield of viral ribonucleic acid (RNA) from nasopharyngeal samples, nonspecific symptomatology, risk of perioperative viral aerosolization, postviral reactive airway with critical respiratory events during extubation, unexpected postop ventilation, and myocarditis (38–50%).[5-7] Data on clinical outcomes in pediatric patients with COVID-19 undergoing emergency surgery is sketchy and mostly extrapolated from adult studies.[5-7] This was a multidisciplinary effort involving cardiac- and neuroanesthesiologists and cardiac and neurosurgeons with perioperative intensive care management for successful outcome.

Goal of perioperative management was to maintain balance between cardiac and cerebral physiology, so that cerebral perfusion was adequate and ICP changes were minimal along with maintaining systemic vascular resistance and optimal cardiac output to prevent intracardiac shunt flow reversal.

Anesthesia was induced, avoiding excess sympathetic stimulation, hypercapnia, and hypoxia to prevent PA pressure and ICP fluctuation.[2,3,8,9] We used sevoflurane (< 1 MAC) and thiopentone for induction. Use of sevoflurane is associated with possibility of raised ICP. However, open fontanelles in this age group and use of sevoflurane in low concentration can seldom cause rise in ICP. Alternatively, IV induction may be a preferred technique. Midazolam and fentanyl, as premedication, alleviated intubation response and surge in ICP; balanced air oxygen mixture maintained cerebral blood flow and metabolic rate.[3,6] Nitrous oxide was avoided as it increases cerebral blood flow, ICP, at higher concentration and reverses protective hypoxic pulmonary vasoconstriction, which is deleterious for causing shunt reversal.[2,8,9] Ventilatory setting was guided by capnography (ETCo2 30–35 mmHg), which maintained normocapnia. Meticulous invasive hemodynamic monitoring, prophylactic inotropes after thoracotomy, and judicious fluid administration with blood transfusion helped to maintain mean arterial pressure (MAP) (45–75 mmHg) and hematocrit (35%). Temperature was monitored and normothermia was maintained as children are prone to hypothermia, because of larger surface area and more so in this child, as thoracotomy was done and expected insensible loses were higher.

Patients with acyanotic heart disease with left-to-right-shunt and severe PAH often present with congestive heart failure and failure to thrive with recurrent chest infection due to increased pulmonary flow.[6] So, ligation of PDA and PAB were performed as a palliation to limit the pulmonary flow.[9]

The rationale behind performing cardiac corrective surgery before shunt placement is open cerebral fontanelles at this age, which compensates for raised ICP.[5] Signs of hydrocephalus were present, but her neurological status permitted the team to perform the shunt placement after the PAB and PDA ligation.

Children with DWM with recent history of COVID-19 infection can potentially present with additional cardiorespiratory complications. To achieve successful postoperative outcomes in such cases, detailed evaluation of central nervous system, cardiorespiratory function, and airway is necessary. The key to manage these cases is to preserve the delicate interplay between cerebral and cardiac physiology.

Conflict of Interest

None declared.

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