Dandy Walker syndrome with VP shunt and laparoscopic surgery- an anesthetic challenge

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INTRODUCTION:

Dandy-Walker syndrome (DWS) is the most common posterior fossa malformation. Classic anatomical hallmarks of DWS are hypoplasia of the cerebellar vermis, enlargement of the posterior fossa and cystic dilatation of the fourth ventricle [1]. The estimated prevalence of a Dandy-Walker malformation is about 1 per 30,000 live births.

Associated congenital anomalies are present in 48% of cases and include orofacial deformities and cleft palate, polydactily and syndactily, cardiac anomalies and urinary tract abnormalities. Central nervous system (CNS) abnormalities include spina biffida, LS meningoceles and agenesis of corpus calosum with poor intellectual development [2].

Development is often affected, particularly with delay in motor skills such as crawling and walking. Spastic paraplegia and seizures may be present.

Many have a sporadic inheritance with some possibly having an autosomal dominant or X-linked inheritance.

Symptoms often occur in early infancy with 80-90% of individual becoming symptomatic in the first year of life. Hydrocephalus, and macrocephaly as its manifestation, is the most common symptom, and DWS accounts for approximately 7.5% of the cases of infantile hydrocephalus. .

The treatment of hydrocephalus involves the shunting of cerebrospinal fluid to another area of the body, and ventriculoperitoneal (VP) shunting is the most common approach.

Laparoscopic approach to surgical procedures had a slower onset in pediatric surgery than in adult patients. But the improvement of laparoscopic surgical techniques has lead to expanded indications for this technology in children

CASE REPORT:

A 9-year-old male (weight 26 kg) was scheduled for elective laparoscopic splenectomy and cholecystectomy. After the birth he was treated for perinatal asphyxia, conatal sepsis, meningitis and cerebral hemorrhage with ventriculomegaly. Dandy-Walker Syndrome was diagnosed and ventriculoperitoneal shunt was placed. He was anemic due to spherocytosis and needed multiple blood transfusions over the years which resulted in splenomegaly and cholelithiasis. He had enuresis and fecal incontinence but no associated heart malformation. Neurosurgeon was consulted prior to surgery to verify the proper functioning of the VP shunt. Standard American Society of Anesthesiologists anesthetic monitoring was used. Pre-medication with midazolam 4 mg PO was given. Anesthesia induction was conducted with propofol, fentanyl and rocuronium bromide. Patent was intubated with endotracheal tube. Tube position was confirmed by capnography and bilateral normal breathing sounds. Anesthesia was maintained with sevoflurane, fentanyl and rocuronium bromide.

Mechanical ventilation was adjusted to keep end tidal CO₂ levels < 35mmHg. Arterial CO₂ was measured: after induction, after insufflation of CO₂ and after 1 hour of operation. Measured arterial CO₂ reflected the values of etCO₂. The shunt reservoir was checked occasionally for over inflation. Surgery lasted approximately 3 and half hours. Vital signs were stable during the operation, which underwent uneventfully. The patient was discharged from the recovery room 2 hours after procedure without any complications.

DISCUSSON:

Anesthetic management of DWS patients is challenging as it can affect several organ systems.

At this time, more abdominal operations in children can be performed laparoscopically.

Pneumoperitoneum results in increased intraabdominal pressure (IAP) and the major concern is that increased IAP could delay drainage, cause shunt malfunction and retrograde leakage of CO₂ into the ventricle, which all may result with increase in intracranial pressure (ICP) [3].

Increased IAP potentially impairs diaphragmatic motion, decreases cardiac output and splanchnic venous return and increases systemic vascular resistance. A significant amount of CO₂ may be absorbed during laparoscopy.

Patient positioning may also impact cardiopulmonary function during laparoscopy and increase ICP.

The main mechanisms for increased ICP are increased intrathoracic pressure and impaired venous drainage rather than increased arterial CO₂ because of systemic CO₂ diffusion [4].

To-date, there are no reports of retrograde shunt failure resulting in pneumocephalus [4].

Concerning to infection risk, the laparoscopy is at least as safe as the open approach in pediatric patients [5].

In two cases complications related to laparoscopic surgery in patients with VP shunts were reported. First patient, undergoing laparoscopic surgery ten days after placement of VP shunt, developed subcutaneous emphysema along the catheter tract. Second patient, with VP shunt placed 5 days before laparoscopic surgery, developed lethargy and apnea.

Therefore, elective laparoscopic cases should be delayed in patients with recently placed VP shunts [3].

Pre-operative evaluation should focus on cardiac, respiratory and neurologic compromise. During anesthesia mechanical ventilation must be adjusted to maintain a physiologic etCO₂ with a goal of <35 mmHg and it is advised to retain insufflation pressure at minimum required and less than 15 mmHg.

Laparoscopic surgery should not be a contraindication in patients with VP shunts, but it must be carried out with good anesthetic and monitoring facilities.

1. Radiopaedia.org/articles, Dandy-Walker malformation, Dr Henry Knipe, A. Prof Frank Gaillard, et al.

2. International Journal of Current Medical And Applied Sciences, volume.3.Issue:1,2014. Anaesthetic Management of an Adolescent with Dandy Walker Malformation. Sanhita Kulkarni et al.

3. Hawaii Med J. 2010 Apr; 69(4): 103–104. Laparoscopic Cholecystectomy in a Patient with a Ventriculoperitoneal Shunt. Chet W Hammill, MD et al.

4. J Gynecol Endosc Surg. 2011 Jul-Dec; 2(2): 91–93. Safety of Laparoscopy in Ventriculoperitoneal Shunt Patients. Rajendra Sankpalet al.

5. Journal of Laparoendoscopic & Advanced Surgical Techniques Vol. 19, No. 5. The Safety of Laparoscopy in Pediatric Patients with Ventriculoperitoneal Shunts. Jason D. Fraser et all.