ANAESTHETIC MANAGEMENT OF A RARE CASE OF 12-YEAR-OLD CHILD WITH CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES WITH A MIGRATED VP SHUNT

Dr. Priya[1], Dr. Kaushik Cherukuri[2], Dr. Arpith Shenava[3*]

1. Junior Resident, Department of Anaesthesiology, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, 411018
2. Senior Resident, Department of Anaesthesiology, Tata Memorial Hospital, Parel, Mumbai, 400012
3. Junior Resident, Department of Anaesthesiology, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune, 411018

[*]Corresponding Author
Dr. Arpith Shenava, 3rd Year Post Graduate, Department of Anaesthesiology, Dr. D. Y. Patil Medical College. Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, 411018, Pune, Maharashtra, India

ABSTRACT

Congenitally Corrected- Transposition of Great Arteries is a rare congenital heart disease where double discordance occurs. It may present with associated cardiac anomalies. Children with CC-TGA get diagnosed usually at an early stage in their lives due to these associated anomalies. We report a diagnosed case of congenitally corrected transposition of great arteries in a 12-year-old boy who presented with abdominal pain with a history of previous surgery for hydrocephalus. On further examination and imaging, it was found that his shunt had migrated to his peritoneum and coiled. He underwent revision ventriculo-peritoneal shunting and laparoscopic removal of the previous shunt from his abdomen.

Keywords: Congenitally corrected transposition of great arteries, migrated VP shunt, congenital heart disease

INTRODUCTION

Congenitally corrected- Transposition of Great Arteries (CC-TGA) is a rare anomaly that occurs in 0.5% of all congenital heart defects[1]. In CC-TGA, both ventricles are reversed along with the main arteries. So, the systemic venous inflow to the right atrium travels to the left atrium and then anomalously to the pulmonary artery. The pulmonary venous inflow enters the left atrium which flows to the right ventricle and then to the aorta[2]. Hence, Atrio-ventricular and ventriculoatrial discordance occurs (double discordance). Although the blood flows in a normal direction, it flows through the wrong ventricles, so the heart actually corrects its deformity.

These patients usually present with an associated anomaly. The three most common anomalies are ventricular septal defect, pulmonary stenosis, and tricuspid regurgitation[3]. The absence of

any early corrective surgery makes the right ventricle the systemic ventricle and can lead to progressive RV dysfunction[4].

CASE REPORT

A 12-year-old boy presented with complaints of pain abdomen for one month. He was previously operated for ventriculo-peritoneal shunt at one year of age because of hydrocephalus. During the workup for the first surgery, he was also diagnosed with congenitally corrected transposition of great arteries. On examination, he was found to be conscious and alert though intellectually challenged. He had no signs of peripheral cyanosis with a heart rate of 96 beats per minute, blood pressure of 112/78 mm of mercury in the right arm in the supine position, saturation of 82-86 percent in all four limbs, and adequate mouth opening. His Glasgow coma scale was 15/15, had a power of four by five in all four limbs. On auscultation, a pan-systolic murmur was heard and air entry was bilaterally equal with normal vesicular breath sounds. On palpation, his abdomen was soft with no tenderness and organomegaly. On further imaging, his electrocardiogram showed sinus tachycardia with a rate of 100 beats/minute and his chest radiograph showed dextrocardia as shown in figure 1. His echocardiography showed (Solitus, L-loop, Levo) with a large malpositioned ventricular septal defect, and severe pulmonary stenosis with good bi-ventricular function. Radiograph of skull and abdomen showed broken VP shunt and migrated VP shunt respectively as shown in figure 2 and 3. His computed tomography of the brain showed asymmetric dilatation of both ventricles suggestive of hydrocephalus and computed tomography of the abdomen and pelvis showed a part of the ventriculo-peritoneal shunt which had migrated and found to be coiled in the pelvis and was confirmed as the cause for his abdominal pain. It was planned to do a revision ventriculo-peritoneal shunt and also a laparoscopic removal of the previous shunt from his abdomen and he was started on propanolol one mg/kg/day by the cardiologist.

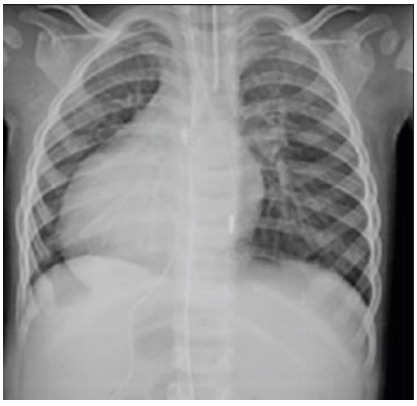


Figure 1

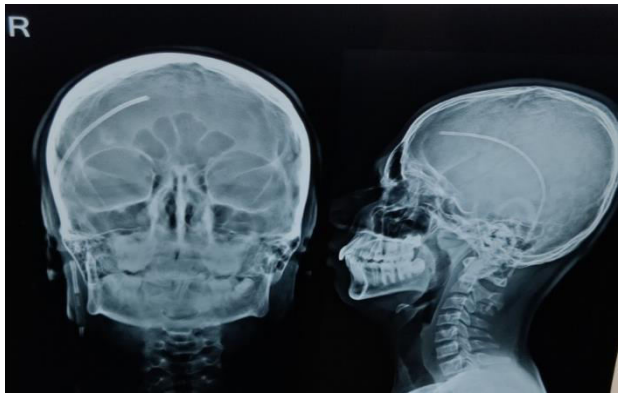
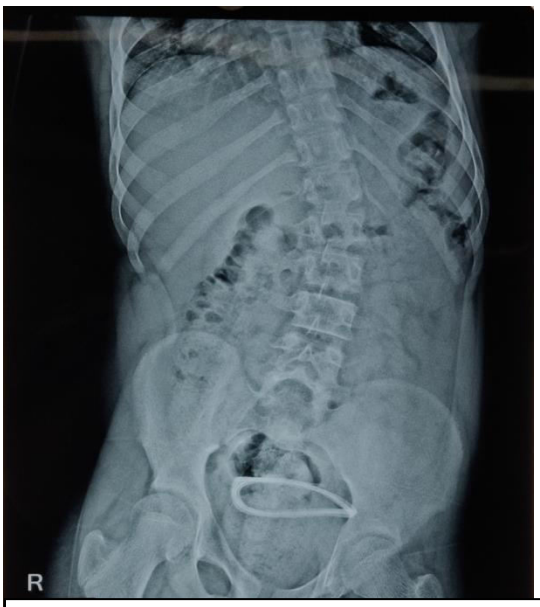


Figure 2



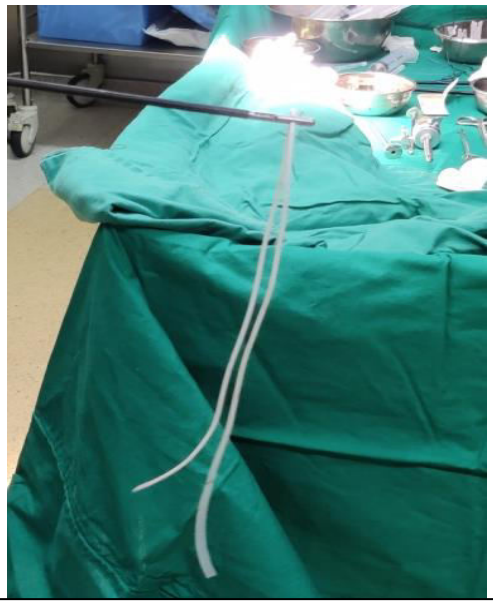


Figure 4

Figure 1: Chest Radiograph of patient with CC-TGA. Figure 2: Skull radiograph. Figure 3: Abdomen x-ray. Figure 4: Removed migrated shunt from abdomen

On the day of surgery, the patient was taken inside the operation theatre and premedicated with intravenous Midazolam two mg and glycopyrrolate 200 micrograms. Injection fentanyl 50 micrograms and ketamine 50 mg were administered along with 10 mg of propofol and a loading dose of 4 mg of intravenous vecuronium. Intubation was performed using a C-MAC video laryngoscope to reduce laryngoscopic stress. A scalp block with 15 ml of injection ropivacaine and 10 ml of injection lignocaine with adrenaline was administered. Intraoperatively, an infusion of 50 mg of ketamine and 50 mcg of dexmedetomidine was started and titrated over the course of surgery. Before skin closure, the infusion was stopped and the patient was extubated uneventfully and shifted to the pediatric ICU for observation.

DISCUSSION

Given the patient's CC-TGA, our anesthetic management was tailored so that we could avoid laryngoscopic stress, sudden hypotension and maintain stable hemodynamics throughout the surgery. A scalp block was given to blunt the response to the incision. The induction was done with a mixture of ketamine and low-dose propofol to avoid hypotension.

Intraoperatively, an infusion of ketamine was started to increase the afterload to divert blood through the pulmonary arteries[5]. Infusion of dexmedetomidine was also started simultaneously to decrease intraoperative opioid usage and for intraoperative hemodynamic stability.

Dexmedetomidine is a centrally acting 2 agonist. A loading dosage of 1 mcg/kg must be infused intravenously during a period of 10 to 15 minutes, followed by an infusion at a rate of 0.2-0.7 mcg/kg/hr[6]. The effects of intravenous and inhalational anesthetics are enhanced by dexmedetomidine. It lowers the heart rate while attenuating the hypertensive hemodynamic response to extubation and intubation by inducing sympatholysis. It also has an opioid-sparing action, reducing the need for inhalational anesthetics and causing a smooth emergence without

any hemodynamic response[7]. It is also postulated that dexmedetomidine decreases the systemic blood pressure, thus decreasing the shunt and hence the pulmonary blood flow and Pulmonary Artery Pressure[8]. Owing to these properties, it can be an adjunct in the anesthesia management of a patient with pulmonary hypertension with general anesthesia.

CONCLUSION

Congenitally corrected transposition of great arteries is a very rare condition and such cases when planned for elective surgery need meticulous anesthetic planning and preparation. The use of ketamine and dexmedetomidine intravenously helped to protect against such complications by increasing afterload and maintaining hemodynamic stability.

The migrated VP shunt being the cause of pain abdomen itself was a rare occurrence but along with co-morbidities like CCTGA, intellectual disability and the patient being of the pediatric age group made this an anesthetic challenge.

REFERENCES

1. Mah, K., & Friedberg, M. K. (2014). Congenitally Corrected Transposition of the Great Arteries. Circulation: Cardiovascular Imaging, 7(5), 849–851.
2. Stanislavsky A. Congenitally corrected transposition of the great arteries (L-TGA) and dextrocardia. Case study, Radiopaedia.org (Accessed on 02 Jan 2023)
3. Chaudhuri, M., & Tomar, M. (2020). Echocardiographic Approach to Congenitally Corrected Transposition. Journal of the Indian Academy of Echocardiography &Amp; Cardiovascular Imaging, 4(3), 312.
4. Obongonyinge, B., Namuyonga, J., Tumwebaze, H. et al. Congenitally corrected transposition of great arteries: a case series of five unoperated African children. J Congenit Heart Dis **4**, 8 (2020).
5. Christ, G., Mundigler, G., Merhaut, C., Zehetgruber, M., Kratochwill, C., Heinz, G., & Siostrzonek, P. (1997). Adverse Cardiovascular Effects of Ketamine Infusion in Patients with Catecholamine-Dependent Heart Failure. Anaesthesia and Intensive Care, 25(3), 255–259.
6. Nair AS. Dexmedetomidine in pulmonary hypertension: A Review. Anaesth Pain & Intensive Care 2013;17(3):279-281
7. Carollo DS, Nossaman BD, Ramadhyani U. Dexmedetomidine: a review of clinical applications. Curr Opin Anaesthesiol. 2008 Aug;21(4):457-61.
8. Kanchi M, Inderbitzin DT, Ramesh KN, Suresh PV, Mayya SS, Sivanandam S, Belani K. Effect of dexmedetomidine on pulmonary artery pressure in children with congenital heart disease and pulmonary hypertension. Ann Card Anaesth. 2020 Oct-Dec;23(4):465-470.