

## Diffuse Cerebral Atrophy, Central Sinus Venous Thrombosis, Hypoplastic Carpus Callosum, Adhd and Vsd in A Child; Anaesthetic Implications

Case Report

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### Abstract

We report a 2.5 year old female child posted for squint surgery of both eyes under general anaesthesia. Child had microcephaly, subtle dysmorphic features, tracheostomy scar, global developmental delay and ADHD. Child also had diffuse cerebral atrophy, hypoplastic carpus callosum, acute on chronic central venous sinus thrombosis partially recanalised, subdural hygroma in left frontoparietal region and ventricular septal defect. ENT opinion was sought to rule out any bands within the trachea because of tracheostomy. Mother was accompanied with the patient in the operation theatre before induction to reduce the anxiety. Inhalational Induction was done in distraction technique with sevoflurane in an incremental manner quickly upto 8vol% via facemask. Direct laryngoscopy was done and trachea was incubated successfully in a second attempt. Anaesthesia was maintained with oxygen, nitrous oxide and sevoflurane. Procedure took three and half hours. Patient tolerated anaesthesia well. Child was extubated after the child was completely awake. Propofol one mg/kg was given ten minutes before extubation to avoid emergence delirium. Intravenous methylprednisolone 10 mg was given intravenously to reduce the edema of the muscles (especially both medial rectus) which might reduce the incidence of bradycardia in postoperative period. Ondasetron 0.1 mg was given slowly i/v in the peri and postoperatively to reduce PONV (Both ADHD and squint surgery has increased risk of emesis). Child was monitored for seizures, bradycardia and other hemodynamic parameters in quiet intensive care environment.

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### Introduction

Central venous sinus thrombosis (CSVT) in neonates, infants, and children is often multifactorial in etiology, with a predisposing comorbid condition or infirmity identified in up to 95% of those affected<sup>1</sup>. These conditions include common childhood illnesses such as fever, infection, dehydration, and anemia, as well as acute and chronic medical conditions such as congenital heart disease, nephrotic syndrome, systemic lupus erythematosus and malignancy [1]. Attention deficit hyperkinetic disorder (ADHD) is the most common neurobehavioral disorder of childhood with an incidence of 5% in school children. It is characterized by inattention, poor impulse control, motor overactivity and restlessness [2]. We report a case of a 2.5 year old female child with diffuse cerebral atrophy, hypoplastic carpus callosum with central sinus ve-

nous thrombosis, ADHD and ventricular septal defect posted for squint surgery of both eyes under general anaesthesia.

### Case Report

A 2.5 year old female child weighing 10 kg posted for squint surgery of both eyes under general anaesthesia. She was born with full term caesarian delivery, cried immediately after birth weighing 2.8kg. Child had microcephaly, subtle dysmorphic features, generalized hypotonia, poor feeding, global developmental delay and failure to thrive. At the age of 8 months, she was diagnosed with congenital heart disease having ventricular septal defect with severe pulmonary arterial hypertension, large post tricuspid shunt and cardiac failure. She was operated for the same 17 months ago. Child had complex postoperative course. She developed seizures. She also developed antibiotic induced enterocolitis treated conservatively. On investigations found to have sepsis and CT scan of brain showed diffuse cerebral atrophy, hypoplastic carpus callosum, acute on chronic central venous sinus thrombosis partially recanalised and subdural hygroma in left frontoparietal region. Child was put on anti epileptic drugs and heparin followed by warfarin. There was difficulty in weaning in postoperative period. Patient was on ventilator for 40 day with tracheostomy and inotropic drugs and in intensive care for almost three months. Tracheostomy scar was closed, gradually condition improved and she was discharged after 98 days.

At present, child was posted for squint surgery (four muscles) of both eyes under general anaesthesia. Blood investigation revealed normal hemogram, renal and liver profile. TORCH profile was negative except Ig G antibodies for CMV was strongly positive suggesting congenital CMV infection and IgM rubella was positive. Ultrasound abdomen was normal. CT Scan of brain showed hypoplastic carpus callosum, diffuse cerebral atrophy, resolving

sub dural hygroma and chronic sinus venous thrombosis with partial recanalisation. ECHO shows small VSD with no pulmonary arterial hypertension and good biventricular function. She was not on any cardiac drugs or warfarin. Child was undergoing behavior and medical (only nutritional) therapy for ADHD. She was seizure free from last 6 months. She was able to walk and speak. ENT opinion was sought and said that the trachea was clear and devoid of any adhesive bands.

Patient was taken as first case with standard fasting protocols. Parent was accompanied with the patient in the operation theatre before induction. All parameters like ECG, NIBP, EtCO<sub>2</sub>, temperature was applied. Inhalational induction was done in distraction technique with sevoflurane in an incremental manner quickly up to 8vol% via facemask. Intravenous line was secured. Intravenous glycopyrrolate 0.06mg/kg, fentanyl 15mic was given. After making sure of adequate ventilation with the bag and mask, atracurium 5 mg was given and ventilated with 100% oxygen for 3 minutes. Direct laryngoscopy was done and trachea was intubated successfully in a second attempt. Intraarterial and central line was secured. Intravenous fluid was given according to CVP to maintain good hydration. Patient was catheterized. All vital parameters were monitored throughout the surgery.

Anaesthesia was maintained with oxygen, nitrous oxide and sevoflurane. Procedure took three and half hours. Patient tolerated anaesthesia well. Child was extubated after the child was completely awake. Propofol one mg/kg was given ten minute before extubation to avoid emergence delirium. Intravenous methylprednisolone 10 mg and ondasetron 0.1 mg was given intravenously. Child was monitored for seizures, bradycardia and other hemodynamic parameters in a quiet intensive care environment for 24 hours. Child had two episodes of vomiting in the postoperative period. She required sedation with midazolam 0.05mg/kg once in the night. She was discharged on third postoperative period.

## Discussion

The incidence of childhood CSVT varies between 0.4 and 0.7 per 100,000 children per year [3] CSVT in infants and children is often multifactorial in etiology. These conditions include common childhood illnesses such as fever, infection, dehydration and anemia, as well as acute and chronic medical conditions such as congenital heart disease, nephrotic syndrome, systemic lupus erythematosus and malignancy [1]. CSVT has also been reported in chronic anemias, such as hemolytic anemia and Evans syndrome [4].

Dehydration and hypovolemia should always be carefully assessed and corrected to prevent thrombus propagation and promote recanalization of the affected vessel [1]. The clinical manifestations of CSVT are nonspecific, may be subtle [1] and may overlap with predisposing conditions such as infection and dehydration [5]. Seizures, altered levels of consciousness and encephalopathy, focal neurologic deficits and diffuse neurologic symptoms (headache, nausea, emesis) may result. CSVT-specific mortality is less than 10%, but motor and cognitive sequelae may require long-term rehabilitative regimens [5]. Coma is a predictor of death in childhood CSVT [5].

ADHD is the most common neurobehavioral disorder of childhood with an incidence of 5% in school children. Characterized by inattention, poor impulse control, motor overactivity and restlessness. Inadequate dopamine and noradrenaline in the fronto-

subcortical-cerebellar regions may cause under stimulation of inhibitory pathways [2]. The effectiveness of stimulants in treatment supports this theory. Poses similar problems to anaesthetists as autistic spectrum of disorders [2].

Perioperatively, children tolerate poorly waiting long periods in hospitals and behavior may become disruptive on the ward, minimize waiting times wherever possible, plan to do the case in the day when the child is more cooperative (ideally, first on a morning list) providing a quiet room to waiting can reduce preoperative anxiety and adverse behavior. Distraction techniques may help when the child is waiting and at induction of anaesthesia. Sedative premedication is used frequently in this group of patients and effect is less predictable and more variable in more variable in patients on stimulants. Medication modifies noradrenergic and dopaminergic functions in CNS. They may reduce the seizure threshold and predispose to PONV. Stimulants like methylphenidate may increase the MAC value of anaesthetic agents [2].

Difficult intubation should be anticipated. ENT opinion should be sought to rule out any bands within the trachea because of tracheostomy. Fibre optic guided intubation can be very helpful. Use of depolarizing relaxant should be restricted in view of malignant hyperthermia and hyperkalemia. Dehydration should be avoided in view of central venous sinus thrombosis. It should be taken as first case and adequate clear liquids can be allowed two hours prior to surgery. Euvolemia should be maintained peri and post operatively according to the central venous pressure. Prophylactic phenytoin can be given to avoid seizures. Heart rate should be monitored over 24 hours for bradycardia as both the medial rectus was operated. Prophylactic steroid should be given to reduce postoperative oedema of operated extraocular muscles and bradycardia. Recovery should be smooth. If sevoflurane is used, emergence delirium can be reduced by giving propofol one mg/kg ten minutes before extubation. Child may require sedation in intensive care because of ADHD. It should be carefully titrated according to the need.

## Conclusion

Central venous sinus thrombosis and ADHD are major issues of anaesthetic concern in our case. Accompanying parents before induction, ruling out any adhesion band in trachea, anticipating difficult intubation, maintaining good hydration according to central venous pressure, reducing emergence delirium by giving propofol if sevoflurane used, monitoring heart rate in postoperative period, adequately preventing and treating PONV, maintaining a quiet and friendly environment in the intensive care and sedating the child according to the need will come a long way in successful management of these type of cases.

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