ANESTHETIC MANAGEMENT OF A CHILD WITH TAUSSIG BING ANOMALY WITH CEREBELLAR ABSCESS POSTED FOR SUBOCCIPITAL CRANIECTOMY

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ABSTRACT
Three and a half year old child diagnosed to have Taussig bing anomaly at 40 days of life now presented with uprolling of eyes and respiratory distress. On admission child was restless, tachypnoeic, had clubbing, cyanosis and low oxygen saturation. He was responding only to painful stimuli. On evaluation computerized tomography brain revealed cerebellar abscess with hydrocephalus causing mass effect and midline shift. Echocardiography showed double outlet right ventricle with subpulmonary ventricular septal defect. He underwent suboccipital craniectomy and abscess drainage under general anesthesia. Post operatively child recovered well without neurological sequelae.

Keywords: Congenital Heart Disease, Taussig Bing Anomaly, Double Outlet Right Ventricle, Cerebellar Abscess

INTRODUCTION
Taussig-Bing anomaly is a rare congenital cardiac malformation that was first described in 1949 by Taussig (1898–1986) and Bing (Konstantinov, 2009). Taussig-Bing anomaly consists of transposition of the aorta to the right ventricle and malposition of the pulmonary artery with subpulmonary ventricular septal defect (VSD) (Taussig and Bing, 1949). They can present in early part of life with symptoms and signs due to shunt, causing cyanosis, clubbing, hypoxemia, polycythemia, volume overload into pulmonary circulation and failure of filtering mechanism in the lungs.

CASES
Three and a half year old boy diagnosed to have Taussig bing anomaly (TBA) at 40 days of life, presented to us with uprolling of eyes and respiratory distress for a day. He was a term child, born by normal vaginal delivery to nonconsanguinous parents with a birth weight of 3.5 kilograms. Child had cyanosis since birth and history of delayed milestones. On admission child was afebrile, restless, tachypnoeic with acidotic breathing, central cyanosis and clubbing. On examination heart rate was 140/minute, blood pressure 90/60 mm Hg, respiratory rate 40/minute with SpO2 of 65% at room air and improved to 80% with 5 litres of oxygen. Child was responding only to painful stimuli with Glasgow coma scale (GCS) of 8/15. Pupils were equal in size of 3mm, reacting to light, bilateral plantar extensor with brisk deep tendon reflexes. Systemic examination revealed precordial prominence, ejection systolic murmur in parasternal area with loud P2 and bilateral conducted sounds in lung fields. Initial resuscitation was done with intra venous fluids and oxygen. Hematological and biochemical investigations were normal except hemoglobin of 18 g/dl. Arterial blood gas analysis showed pH 7.46 PaCO2 29.5 mmHg PaO2 35mmHg HCO3 20.3 mmHg lactate 5.4 mmol/litre and SaO2 80% with fractional inspired oxygen concentration of 0.3. Chest X-ray showed cardiomegaly and pulmonary plethora. Neurosurgeon opined possibility of brain abscess. Computerized tomography (CT) brain showed left sided cerebellar lesion probably abscess with hydrocephalus causing mass effect and midline shift. Anti cerebral edema measures and antiepileptics were started. Planned for emergency left sided suboccipital craniectomy with drainage of abscess. The cardiologist was consulted and echocardiography showed situs solitus, levocardia, intact interatrial septum, double outlet right ventricle, large subpulmonary VSD with bidirectional shunt and severe pulmonary arterial hypertension. Meanwhile elective mechanical ventilation initiated prior to surgery in view of poor GCS. Triple lumen central venous cannula was

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inserted through right internal jugular vein. Informed high risk consent obtained for general anesthesia explaining perioperative cardiac events. Our anesthetic concerns were patient in paediatric age group with cyanotic congenital heart disease and single ventricular physiology posted for major neurological procedure in prone position. The anesthetic goals were to maintain adequate hydration, to avoid hypoxia, acidosis, increase in pulmonary artery pressure and decrease in systemic vascular resistance. Infective endocarditis prophylaxis was also given. Child shifted to operating room, connected to standard American society of anesthesiolgist (ASA) monitors, anesthesia was induced and maintained with injection fentanyl, vecuronium, sevoflurane, oxygen and air. Child placed in prone position, left sub occipital craniectomy and abscess drainage was done uneventfully. Post surgery child was electively ventilated in view of poor pre operative GCS. Post operative CT scan brain showed reduction in abscess cavity size and hydrocephalus. Child had subtle seizures on post op day1, responded to step up dose of antiepileptics. Child weaned off ventilator on day 2 and extubated with no neurological sequelae. Discharged subsequently. Child could not undergo corrective surgery for taussig bing anomaly due to financial constrains. On one year follow up child had right and left hemiparsis on two occasions due to thrombosis of cerebral vessels and cerebral infarction but gradually recovered well.

Figure 1: Chest X-ray showing cardiomegaly  
Figure 2: CT brain showing cerebellar abscess

DISCUSSION
Taussig-Bing anomaly is the second most common type of double outlet right ventricle (DORV) (Walters et al., 2000). DORV is a group of abnormal ventriculo-arterial connections where both pulmonary artery and aorta arise from the morphologically right ventricle. It is a rare condition, affecting 1-1.5% of the patients with congenital heart diseases and occurring in 1 out of 10,000 live births (Silka, 1998). The anatomical types are classified according to the relationship between the ventricular septal defect (VSD) and the blood vessels, the position of the great arteries in relation to each other and the presence of additional malformations. According to the relationship between the VSD and the great arteries, DORV can be classified as DORV with subaortic VSD, DORV with sub pulmonary VSD, DORV with doubly-committed VSD and DORV with non-committed VSD (Peixoto et al., 1999), Taussig-Bing anomaly consists of transposition of the aorta to the right ventricle and malposition of the pulmonary artery with sub pulmonary VSD (Taussig and Bing, 1949). Echocardiography is an useful method for diagnosing this complex malformation. This test accurately identifies the anatomical variables and guides the choice of the most appropriate surgical approach (Peixoto et al., 1999). As aorta and pulmonary artery arises from right ventricle and contains the deoxygenated blood, it has been asserted that a ventricular septal defect is universally present in DORV and that its presence is mandatory for the survival of the patient (Neufeld et al., 1961). Our patient presented with cyanosis, clubbing, hypoxemia, polycythemia, respiratory distress,
severe pulmonary arterial hypertension with bidirectional shunt. Failure of filtering mechanism in the lungs could have resulted in microorganism reaching the systemic circulation that could have caused brain abscess in our patient like in any other congenital cyanotic heart disease. In a study involving twenty-eight patients of cyanotic congenital heart disease (CHD) complicated with brain abscess, DORV with VSD comprised 25% of the cardiac lesions in causing brain abscess (Chakraborty et al., 1989). As with other CHDs, the goal of anesthetic management was to maintain the optimal balance of pulmonary and systemic blood flow despite the surgical stress imposed on the patient. The aim was to prevent peripheral vasodilation or pulmonary hypertension that would increase the right to left intracardiac shunt. Typically, this aim is achieved by minimizing cyanosis with supplemental oxygen and avoiding acidosis and hypercapnia with adequate ventilation and euvolesmia (McClung et al., 2011). Anatomic repair, with connection of the morphologically left ventricle to the aorta and the morphologically right ventricle to the pulmonary artery is the treatment of choice. While this can be achieved with interventricular repair in selected cases (Serraf et al., 1991), the arterial switch operation (ASO) has become the preferred management strategy in many major centers (Rodefeld et al., 2007). Over the last two decades one-year survival in patients after anatomic repair increased from 47% to 100% and the 5-year event-free survival rates increased from 35% to 87% (Alsoufi et al., 2008).

Conclusion
We are presenting this case of Taussig bing anomaly due its rarity, early diagnosis of the disease, its complications and appropriate timely intervention is needed in management of such rare disease to have a good outcome.

REFERENCES