

Management of a Critical Case of Double Outlet Right Ventricle (DORV) and Cerebral Abscess by Multiple Interventions

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Abstract

Tetralogy of Fallot (TOF) is the commonest type of cyanotic congenital heart disease which accounts for 10% of all congenital heart disease. Delay in surgical treatment leads to polycythaemia, cerebral abscess, thrombotic episodes etc. Corrective surgery in a case with cerebral abscess always has risk of intracranial hemorrhage during bypass¹. We are hereby reporting a case of TOF who had multiple cerebral abscess managed with burr hole operation and extra ventricular drainage. Later 5 coronary stents were placed in Right Ventricular Outflow Tract (RVOT) to Main Pulmonary Artery (MPA) to overcome infundibular and valvular stenosis and thus reducing right to left shunt and cyanosis. This is the first ever palliation with RVOT stenting in a case of Double Outlet Right Ventricle (DORV), Ventricular Septal Defect (VSD), Pulmonary Stenosis (PS) with cerebral abscess where surgery was contraindicated at that time and patient condition was unstable. Later on she had bidirectional Glenn shunt on 8th December 2015 by Saudi charity team.

Key-words: Tetralogy of Fallot (TOF), Right Ventricular Outflow Tract (RVOT) Stenting, Cerebral abscess.

Case Report

M, 3 years old baby girl, presented to department of pediatric cardiology, combined military hospital Dhaka on 18.08.14 with the complaints of fever, headache, reluctant to feed and projectile vomiting for one month duration. She was diagnosed as a case of DORV with VSD with PS at the age of one year in another hospital but because of poverty could not avail operative treatment. She had history

of repeated cyanotic spell since 4 months of age which could not be addressed properly in village home at any time. In CMH, Dhaka, she was investigated and echocardiography revealed large mal aligned VSD, severe infundibular and valvular stenosis and 80% overriding of aorta over right ventricle. ECG showed right axis deviation and right ventricular hypertrophy. X-Ray chest showed right ventricular hypertrophy, uplifted apex and oligemic lung fields. Hematological examinations showed raised hemoglobin and hematocrit (60%) level with neutrophilic leukocytosis. Coagulation profile was normal and blood culture was negative. CT scan of brain was performed urgently and multiple cerebral abscesses were identified.

On examination patient was conscious, ill looking, febrile (Temp 103°F), pulse 100/min, BP 90/60 mmHg, clubbing, cyanosis and conjunctival congestion was present. Cardiovascular examination revealed parasternal heave and pan systolic murmur. Neurological examination revealed sign of meningeal irritation.

Treatment was started immediately with injection Meropenem, Injection Phenobarbitone, Tab. Propranolol and other supportive medicines. Injection Morphine was advised for cyanotic spell. Injection Mannitol was added for cerebral oedema. Patient was referred to neurosurgeon immediately for drainage of abscess under general anesthesia. But due to critical condition of patient, she was unfit for general anaesthesia. So surgery was delayed for 10 days. On 28.08.14, Burr hole operation was done and antibiotics were changed to injection Megacillin, injection Linezolid and injection Metronidazole. Patient was febrile and toxic and CT scan repeated

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on 29.08.14 showed ventriculitis with hydrocephalus with left sided multiple abscesses. Extraventricular drainage given on right side on 02.09.14 and left side on 10.9.14. Patient's condition was deteriorating, She developed low output state, hypotension, urinary retention (neurogenic bladder) and fever. On 10.9.14 antibiotics were changed to injection Clindamycin and Cefepime and injection Dopamine and Dobutamine were added.

Fever was persisting up to 08.10.14. Antibiotics were continued for four weeks. On 26.10.2014 the case was referred to a charity cardiac team from King Faisal Specialist Hospital Jeddah who was visiting CMH Dhaka for a charity mission on Paediatric cardiac interventions and surgery. They reviewed her and considering her SPO₂ (45%), and hematocrit (60%) they agreed for an urgent intervention. As she was a high risk case at that time to go for cardio pulmonary bypass and unable to do surgery by own cost from a developed centre abroad, team decided for stenting her RV outflow tract with peripheral stent.

Procedure

Patient was sedated with injection ketamine and draped as usual. Arterial line was established. Right femoral vein was cannulated with 6F sheath. Right ventriculography showed severe infundibular and valvular stenosis. RVOT was dilated with 4x10 mm PTCA balloon. RVOT stenting was done with 4x24, 4x18, 4x18, 4x18 mm stent. Stent covered a length from RVOT to main pulmonary artery. Injection Enoxeparin was injected subcutaneously 1mg/kg after procedure. Second dose was repeated after 12 hours. Tablet Aspirin was advised for 6 months. Patient's condition was improved dramatically after the procedure. Her saturation improved to 85-90%, right to left shunt diminished. She started eating, walking and playing from 3rd day after procedure (She was bed ridden for 3 months and developed bed sore also). She was discharged 7 days after intervention.

Discussion

Brain abscess is a rare but extremely dangerous variety of suppurative disease². Solitary brain abscess constitute only 0.5% to 6% of the reported cases of brain abscess³. The incidence of brain abscess in the population with cyanotic congenital heart disease (CCHD) varies from 5 to 18.5%⁴.

Another study showed CCHD as important pre-disposing factor for cerebral abscess in 25 – 46% of cases⁵. Despite improvement in early suspicion, CT scan facility, medical and surgical management with newly developed antibiotics, the mortality rate for cerebral abscess is still present (13%)⁶. Tetralogy of Fallot (TOF) is the most common cyanotic CHD associated with intracranial suppuration^{7,8,9}. The abscess are most often supratentorial. Only 4% of patients had posterior fossa abscesses in a series of 75 cardiogenic abscess⁷. The prognosis of cardiogenic abscess is worse than that of other brain abscesses. Mortality ranged from 27.5% to 71%⁷. Non hemolytic streptococci are recognized as causative organism of cerebral abscess in most reported cases¹⁰.

Factor responsible in the pathogenesis of brain abscess in patient with cyanotic CHD are, right to left shunting of blood bypasses phagocytic activity of lungs which allow direct entry of blood to cerebral circulation⁹. Low perfusion area in brain resulting from polycythemia leads to tissue hypoxia and acidosis^{11,4}. Microorganism with shunted blood seeded in such area leads to cerebral abscess. The greatest peri-operative concern is the development of cyanotic spells due to spasm of the hypertrophied infundibulum. Tachycardia and increased myocardial contractility can lead to infundibular spasm. Another mechanism of spell is decreased SVR and increased right to left shunt through VSD. Systemic blood pressure less than 60 mmHg can trigger hyper cyanotic spell. Anaesthesia management of TOF cases during brain abscess drainage needs understanding about drugs which can alter magnitude of right to left shunt. Major challenge is to maintain SVR, reducing PVR and provide mild myocardial depression^{11,12}.

Our patient had burr hole operation on 26th August 2014. Eighteen ml thick pus removed. On 28th August repeat CT scan showed ventriculitis and hydrocephalus. Frontal extra ventricular drainage was given on right side. After 7 days CT scan brain showed left sided ventriculitis and hydrocephalus, so left sided EVD was performed. Patient was still febrile and Clindamycin and Cefepime given for 4 weeks. Her condition was still critical so she was referred to cardiac surgeon for corrective surgery. Considering her condition and risk, palliation with right ventricular outflow tract stenting was advised.

Traditional approach to patients with narrow right ventricular outflow tract (RVOT) and reduced pulmonary blood flow seen classically in Tetralogy of fallot (TOF), Double outlet right ventricle (DORV) with TOF physiology are surgical correction. Although surgical skill for BT shunt and total corrective surgery has been improved but still mortality is high in high risk patients. As this case was still in the convalescence period after brain abscess surgery and had low oxygen saturation (40-50%) RVOT stenting was planned.

Aim was to allow more pulmonary blood flow and to reduce right to left shunting through VSD^{13,14}. Premounted coronary stent were selected as peripheral stent of appropriate size was not available in the stock. Her RVOT diameter in systole was 2.5 mm and in diastole was 3.5 mm. Four coronary stents were placed from RVOT to main pulmonary artery and on RVOT was created with 4x24, 4x18, 4x18, 4x18 stent. Patient condition improved dramatically after procedure. Our plan was to go for total correction 6 months to one year after stenting once patient's general condition will improve. After 14 months Saudi charity team arrived and she was presented for surgical intervention.

Considering her right ventricular situation (severe hypertrophy with multiple stent in RVOT), she was accepted for bidirectional Glenn shunt (BDG), which was performed on 8th December 2015. Bidirectional glenn shunt is a communication between superior vena cava and right pulmonary artery by which venous blood from upper half of body is diverted to lungs directly bypassing heart. These kinds of procedure or palliation are performed mainly for single ventricle type of heart to prevent mixing of venous blood with oxygenated blood and to channel venous blood directly to lungs.



Fig-1: Baby M after treatment.

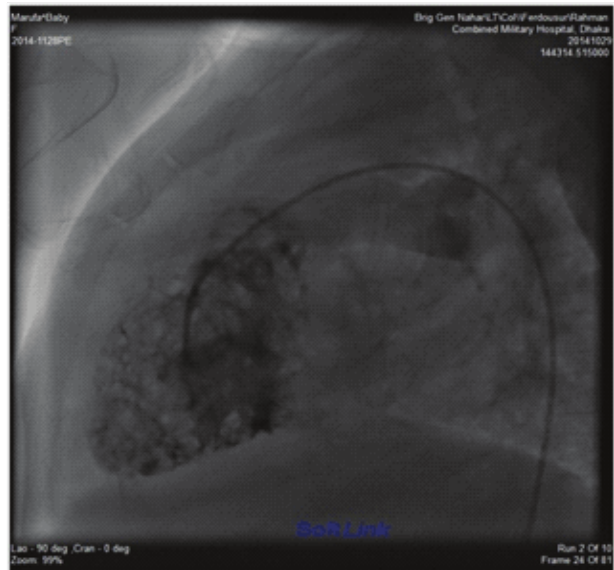


Fig-2: Right Ventriculography showing severe infundibular stenosis.

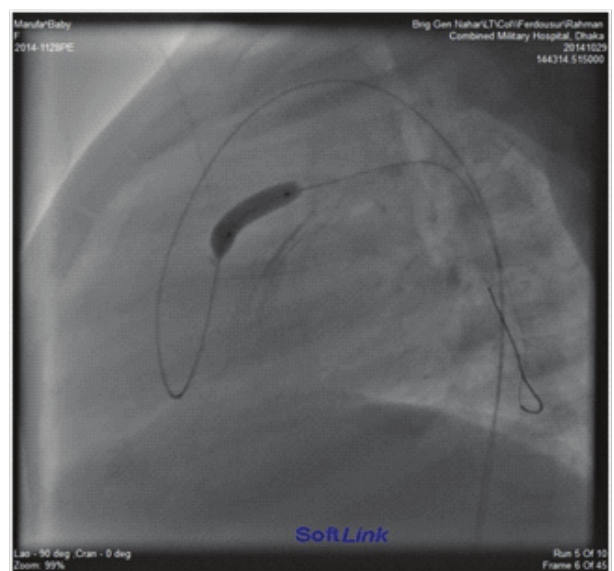


Fig-3: Right ventriculography showing RVOT stent in position.

Conclusion

Peri-operative management of the patient in intensive care unit, operative management by neurosurgery team and RVOT stenting by pediatric cardiac team saved life of this critically ill baby girl who was admitted in hospital for long 3 months and discharged in a satisfactory condition with oxygen saturation of more than 85%. Team work of all concerned sub specially landed to a good outcome for this child.

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