Anaesthetic Management Of Pulmonary Atresia Posted For Non Cardiac Surgery

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Citation

D Singh, P Jindal, G Chopra. *Anaesthetic Management Of Pulmonary Atresia Posted For Non Cardiac Surgery*. The Internet Journal of Anesthesiology. 2005 Volume 11 Number 1.

Abstract

A young male presented for open reduction of fracture femur. On preanaesthetic checkup he was found to be having continuous murmur best heard at 2nd left intercostal space in mid clavicular line. Echocardiography and cardiac catheterisation confirmed the diagnosis to be pulmonary atresia, large ventricular septal defect with overriding of great vessels, large aorto-pulmonary collaterals and patent ductus arteriosus to left pulmonary artery.

CASE REPORT

A 29 years old patient of 55 kg body weight with fracture of the femur was posted for open reduction and internal fixation. The patient had a history of dyspnoea on exertion (NYHA-II), easy fatigability and palpitation since childhood. These symptoms had increased in severity over last three years. There were no history of cough, syncopal attack, orthopnoea, paroxysmal nocturnal dyspnoea, pedal edema and ascites.

On examination, his pulse rate was 80/min regular, fair volume with normal character. There were cyanosis and mild clubbing. There were no evidence of pallor, lymphadenopathy and jaundice. JVP was not raised. Respiratory system examination was within normal limits. Precordial examination revealed cardiomegaly with normal first heart sound. Second heart sound was single and loud. There was continuous murmur which was best heard at 2nd left intercostal space in mid clavicular line. Bilateral Interscapular auscultation also revealed continuous murmur.

Hematological investigations showed-Hb-18.9gm/dl, TLC-7700/mm3, DLC (P-66, L-32, E-02, M-0,), prothrombin time-12 seconds over the control of 12 seconds, platelet count - 1.29/cumm; bleeding time - 1.10 minutes and clotting time was 6.10minutes. Biochemical investigations showed blood sugar-90 mg/dl, blood urea-20mg/dl, serum creatinine-0.9mg/dl, serum sodium- 148mEq/lt, serum potassium-3.9mEq/lt and serum calcium-110mEq/lt. Electrocardiogram showed normal sinus rhythm and right ventricular hypertrophy. Chest x- ray showed normal cardiac- thoracic ratio and oligemic lung field.

He was suspected of having congenital cyanotic heart disease. Patient was advised to undergo echocardiography and cardiac catheterisation.

Echocardiography showed normal mitral leaflets, EF slope was 98.4cm/sec and De amplitude was 26.6mm at mitral valve. Aortic valve was normal with aortic root diameter of 45.2mm and cusp separation of 27.1mm. Tricuspid valve was normal. Pulmonary valve was not visualized. Left ventricle size in diastole and systole was 60.5mm and 37.5mm respectively. Ejection fraction was 58%.Right ventricle size was 20.7mm. Right atrium and pericardium was normal. 2D analysis showed large left ventricle and right ventricular hypertrophy. There was subaortic ventricular septal defect (VSD) of 1.3 cm size with overriding of great vessels which were dilated. The pulmonary valves and pulmonary artery could not be visualized. Interatrial septum was complete. Tissue Doppler analysis showed normal study.

Flow indices showed peak velocity at mitral, aortic and tricuspid valve were 88cm/sec, 114cm/sec and 71cm/sec respectively. There was bidirectional flow at VSD. Trace tricuspid regurgitation was also seen. In suprasternal view multiple vessels with continuous flow were seen.

Cardiac catheterisation revealed pulmonary atresia, right aortic arch, and patent ductus arteriosus to left pulmonary artery. There were large collaterals from arch supplying right lung. SpO_2 of blood in various chambers of the heart was not

reported.

Patient was diagnosed to be having pulmonary atresia with discontinuous pulmonary artery, large ventricular septal defect with overriding of great vessels, large aorto-pulmonary collaterals and patent ductus arteriosus to left pulmonary artery.

ANAESTHETIC MANAGEMENT

The patient was premedicated with tab diazepam (5 mg) and tab ranitidine (150mg) orally in night and morning respectively. Invasive blood pressure, pulse oximetry, electrocardiogram (ECG), end tidal carbon dioxide ($ETCO_2$), and urine output monitoring were done. After securing intravenous line with 16G canula and adequate pre oxygenation, patient was induced with inj thiopentone (300 mg) i.v., intubated after giving inj vecuronium bromide (0.1mg/kg) i.v. and 8.0 no. cuffed oral endotracheal tube was put. Anaesthesia was maintained with 33% O_2 in air, isoflurane and inj fentanyl. At the time of induction and intubation pulse rate was 70-80 per min, blood pressure was110/80 mmHg, SpO_2 was 97% and $ETCO_2$ was 35-40 mm Hg. Special precautions were taken to avoid hypoxia, hypercarbia, hypovolemia, hyperventilation, sympathetic stimulation and hypothermia as these factors precipitate pulmonary hypertension leading to fall in pulmonary blood flow. Utmost care was taken to avoid introducing any air bubble to avoid systemic air embolism (paradoxical embolism). For intra operative and post operative pain, non steroidal anti inflammatory drugs (NSAID) were avoided as it may lead to the closure of patent ductus arteriosus. Throughout the surgery patient remained haemodynamically stable.

Considering the criteria for extubation anaesthesia was reversed with inj. neostigmine and inj. glycopyrrolate. Patient was put on O_2 supplement for 4-6 hrs after extubation in post anaesthesia care unit. Haemodynamically patient was stable and shifted to ward from where he was discharged after a month with advise of having corrective surgery for congenital heart disease in future..

DISCUSSION

Pulmonary atresia is an uncommon condition affecting 1% to 3% of patients with congenital heart disease and is a frequent cause of death in neonatal period.[1]This was first reported in 1783 by Hunter. It is thought to be familial and multifactorially determined, although a recent report suggests an autosomal recessive inheritance.[2]

Embryologically the semilunar valves are formed from small tubercles found on the main truncus swelling. They are visible when the truncus partitioning is nearly complete. Two pairs of these tubercles form the pulmonary and aortic valves. These tubercles gradually hollow out on their upper surfaces, thereby forming the semilunar valves.

In pulmonary atresia the valves remain fused and may form a dome shaped structure. The pulmonary trunk may be narrow or atretic and the only outlet from the right heart will then be a patent foramen ovale. A patent ductus arteriosus form the path of blood to the pulmonary circulation.

Pathophysiologically the fusion of the pulmonary valve cusps into a membrane that does not allow blood to exit from right ventricle in normal manner is the basic abnormality. Here venous return is shunted through ventricular septal defect from right ventricle to left ventricle. A patent ductus arteriosus allows some blood to flow through the pulmonary vascular bed. This is the most frequent type of disorder and associated with a small hypertrophied right ventricle, oligemic lung fields and cyanosis. Survival will depend on the patency of the patent ductus arteriosus and the presence of the ventricular septal defect.

Clinically patients show early severe cyanosis and tachypnoea. There is cardiomegaly and single second heart sound. On auscultation there is a pan systolic murmur of VSD and machinery murmur of patent ductus arteriosus. With progressive right ventricular failure, hepatomegaly and prominent “a” wave of jugular venous pulse may be observed.

On chest X- ray heart size may be normal or enlarged with oligemic lung fields. Right atrium is dilated in the presence of tricuspid regurgitation. Electrocardiogram may show right ventricular hypertrophy. Echocardiograph may show atretic pulmonary valves, diminished tricuspid valve excursion, decreased size of right ventricle and normal or small left ventricle. The condition can be diagnosed during fetal life. [3]

The following anaesthetic management priorities should be considered:

- Appropriate premedication
- Maintaining open patent ductous arteriosus (P D A)
- No rise in pulmonary vascular resistance

- Meticulous purging of all air from intravenous lines

Premedication can be done with tab diazepam in the dose of 0.5 mg /kg body weight, i.m. midazolam in the dose of 0.08 mg/kg body weight or i.m. morphine 0.1 to 0.2 mg/kg body weight. Crying and agitation induced by the pain of an intramuscular injection will aggravate shunting, hypoxemia and acidosis and is therefore best avoided. All sedated patients require monitoring.

Closure of PDA will lead to the demise of patient. This ductus arteriosus can be maintained patent by prostaglandin E infusion at the rate of 0.03 to 0.1 µg/kg/min Succinylcholine is avoided for intubation as acetylcholine constricts the PDA and analgesic like NSAID should not be used.

Pulmonary vascular resistance (PVR) is crucial consideration in this condition. Pulmonary vasculature is highly reactive and any rise in PVR may cause significant fall in pulmonary perfusion and therefore hypoxemia. The conditions that increase PVR are hypoxia, hypercarbia, hypovolemia, hyperacidemia, hyperinflation of lungs (PEEP), hypothermia, sympathetic stimulation, atelectasis and high hematocrit, so should be avoided .[4] Studies of ketamine and nitrous oxide in patient with normal or elevated PVR have shown no increase in resistance provided ventilation is controlled.[5]

De-airing of intravenous line is important consideration as these patients with anatomical shunt is at the risk of paradoxical embolism. The solubility coefficient of a gas in a liquid is temperature dependent. With the warming of fluid bubbles may form as gas escapes from the fluid. Therefore fluid should be prewarmed before transfusing.

CONCLUSION

Pulmonary atresia with ventricular septal defect and patent ductus arteriosus imposes a challenge in cardiac as well as non cardiac surgery. It needs to be investigated properly and managed by avoiding the factors increasing the right to left shunt and air bubble going in the veins causing paradoxical embolism.

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References

1. Freedom RM, Wilson G, Trusler GA et al: Pulmonary atresia and intact ventricular septum: a review of the anatomy, myocardium and factors influencing the right ventricular growth and guidelines for surgical intervention. Scand J Thorac Cardiovasc Surg 1983; 17:1.
2. Erikson NL, Butino L, Jubreg R: Congenital pulmonary atresia with intact ventricular septum, tricuspid insufficiency, and PDA in two sibs, Am J Med Genet.1989; 32:187.
3. Todros T, Gaglioti P, Demarie D: Pulmonary stenosis with intact ventricular septum. Documentation of development of the lesion echocardiographically during fetal life. Int J Cardiol 1988; 19:355.
4. Berry FA: Anaesthetic management of difficult and routine paediatric patients. Ed 2, New York, Churchill-Livingstone, 1990.
5. Hickey PR, Hanson DD, Strafford M et al: Pulmonary and systemic effects of nitrous oxide in infants with normal and elevated PVR. Anaesthesiology.1986; 65:374.

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