

CASE REPORT

Total Abdominal Hysterectomy in a Patient with Uncorrected Truncus Arteriosus

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Introduction

Uncorrected complex congenital cyanotic heart diseases in adults are rare today. Truncus arteriosus is one such rare condition in which only one vessel originates from the heart and from this common vessel, systemic, pulmonary, and coronary circulation originates. Uncorrected only 20 % survive first year of life. We report a case of total abdominal hysterectomy for fibroid uterus done in a patient with uncorrected truncus arteriosus-type 1V .

Case Report

Miss VRF 27 years old, unmarried female, came with a history of 2 months amenorrhoea. Patient is a known case

of complex congenital cyanotic heart disease truncus arteriosus. On Examination: thin built, young female, no Pallor, Cyanosis or Clubbing was proceed, Pulse—70beats/min; BP—90/60 mm of Hg; R.R—18/min, R/S—air entry spual on both side, No added sounds CVS—Pan Systolic Murmur +ve; P/A—16 wks firm non-tender mass arising from the pelvis; On Investigation: Hb—18 gm%; PCV—45 %, BT/CT—wnl; UPT—neg; Bld. Gr.—O+ve; ABG-pH—7.423, Pco2—33, Po2—39.8, Na-142.1, K—4.4.

Ultrasound

Uterus A/V, normal in size, displaced to left by a solid mildly vascular pelvic mass measuring 12 cm × 11 cm × 7.2 cm. Uterine cavity central, ET = 10 mm Rt. ovarian tumor.

CT Scan

Large 17 cm × 11 cm × 8 cm lobulated hyper vascular solid mass arising from pelvis similar to uterine echo texture contiguous with uterine myometrium posteriorly ? fibroid. Both ovaries are normal. Effect of mass is seen on distal ureters with mild bilateral hydronephrosis (Figs. 1, 2).

Cardiac Catheterization Report

Congenital heart disease; fallots tetralogy; pulmonary atresia, large sub aortic VSD; multiple bilateral MAPCAS

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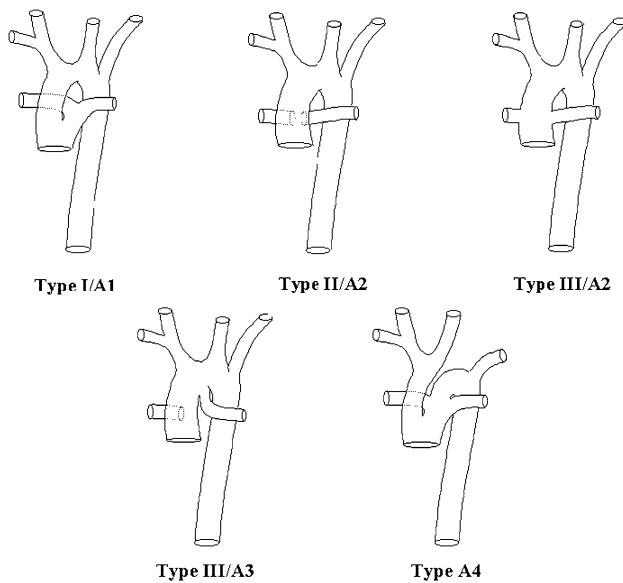


Fig. 1 Types of truncus arteriosus—Sketch diagrams

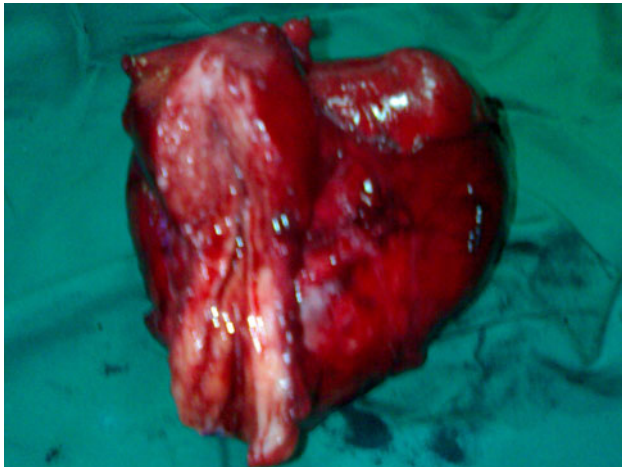


Fig. 2 Post operative picture showing specimen of uterus with fibroid (Cut specimen)

(aorto-pulmonary collaterals) Opn: Type IV Truncus Arteriosus. *Echocardiography*: truncus arteriosus (TYPE B).

In view of the large mass effect caused by the fibroid leading to bilateral hydro-nephrosis the case needed a surgical management for the fibroid. As the patient had a complex congenital cyanotic heart disease-truncus arteriosus, a myomectomy with its accompanying blood loss was not found to be the treatment of choice. Opinion of a cardiothoracic surgeon was taken with regard to future child bearing which was thought to be dangerous with the existing heart condition. After a prolonged discussion with the family and the patient a decision for total abdominal hysterectomy was taken. Patient was a high-risk case for anesthesia [1, 2], ASA Grade-IV Patient was taken up for total abdominal hysterectomy. Rt. Int. Jugular Vein was cannulated with

double lumen arrow central line under LA; Induction done with Preoxygenation, i.v. glycopyrrolate 0.2 mg, i.v. ketamine 60 mg, i.v. vecuronium 3 mg. Oral intubation and controlled ventilation convergent with oxygen (100 %) + seroflurane (0.25 %) for maintenance, with vancuronium top up doses. Sedation was achieved with i.v. pentazocine 15 mg, i.v. midazolam 2.5 mg. Prior to induction Oxygen saturation on air was 78%, Intra operative saturation maintained at 80–86 % (with 100 % oxygen).

On opening the abdomen with Pfannensteil Incision, large postero-right lateral wall sub-serous fibroid noted, tightly fitting in pelvic cavity. Rt rectus abdominus was cut just above its insertion to symphysis pubis to deliver the fibroid uterus out of abdominal cavity. Total abdominal hysterectomy was convergent. Post operatively both ureters showed peristalsis. Abdomen was closed leaving a drain in peritoneal cavity. Reversal was done with i.v. glycopyrrolate and neostigmine, followed by extubation. Post extubation was 80 % on oxygen supplementation. Post operatively patient was maintained on mask oxygenation at 8liters/min which was gradually decreased over a week. Patient had an uneventful recovery and was discharged on 9th post-operative day.

Discussion

Complex congenital cyanotic heart diseases are diagnosed in childhood and in modern era undergo successful surgical corrections. Uncorrected congenital cyanotic heart diseases are rare in adults due to their poor prognosis. Our patient, a twin was a “blue baby” at birth, the other twin a male, is normal. At the age of 3 months, she was diagnosed as Fallot’s Tetralogy surgical correction which was refused by the parents due to high mortality involved. Patient had cyanotic spells intermittently which were managed conservatively. Cardiac catheterization study done in 2004 made a definitive diagnosis of type IV truncus arteriosus.

Path-physiology of truncus arteriosus is typified by cyanosis and systemic ventricular volume overload. Out-flow from both ventricles is directed into the common arterial trunk. Pulmonary blood flow is derived from this combined ventricular output, and its magnitude depends on the ratio of resistances to flow in the pulmonary and systemic vascular beds. Because of the mixing (although not complete) of left and right ventricular output that occurs primarily during systole and at the level of the common arterial trunk, subnormal systemic arterial oxygen saturation is common. Similarly, because the systemic and pulmonary circulations are essentially in parallel, pulmonary blood flow typically is at least 3-fold higher than systemic blood flow, with pulmonary over circulation and increased myocardial work that results in increased resting oxygen

demand and decreased metabolic reserve. The median age of death without surgery ranges from 2 weeks to 3 months, with almost 100 % mortality by age 1 year. Cause of death in un-repaired patients is usually cardiac arrest or multiple organ failure in the face of systemic perfusion that is inadequate to meet the body's metabolic demand, progressive metabolic acidosis and myocardial dysfunction after extensive search no recorded cases of hysterectomy done in patient having truncus arteriosus was found.

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