Anticipate and Prepare. Patient with repaired TOF. From Cardiac Catheterization Procedure to an Open-Heart Surgery

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1. Title of Medically Challenging Case Report Poster:
"Anticipate and Prepare. Patient with repaired TOF. From Cardiac Catheterization Procedure to an Open-Heart surgery".

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3. Learning track/ topic: Cardiac Anesthesia.

4. Learning Objectives:
- To review long term complications of patients s/p Tetralogy of Fallot (TOF) repair.
- To present complications associated with stent migration in the cath lab and to discuss management. Key takeaway: Cardiac catheterization in patients with congenital anomalies is associated with significant morbidity. The anesthesia team needs to be prepared to transition from a minimally invasive to an open surgical procedure.

5. Introduction:
The population of patients with congenital heart abnormalities has been growing. Approximately 85% of babies born with cardiovascular anomalies can expect to reach adulthood (1). There is an increase not only in the survival but also in the birth prevalence of congenital heart diseases. (pic 1) (2)

(Insert Image)

TOF consists of pulmonary stenosis, VSD, overriding aorta and RV hypertrophy. The birth prevalence of TOF is about 0.5/1000, which represents approximately 6% of all newborns with congenital heart disease. This condition is typically managed with an open-heart surgery during the 1st year of the life with correction of RV outflow track and VSD closure. Long term, these patients often require numerous interventions to address pulmonary valve incompetence or diminished right
ventricular outflow tract (RVOT). Over the past decade, minimally invasive techniques are increasingly utilized (>2x) to address inadequacies of the pulmonary valve and pulmonary arteries. (3)

6. Case description:

“A 16-year-old Male s/p TOF repair later PA stent placement had a scheduled PA revision stent placement and a Melody Transcatheter Pulmonary Valve (TPV) implantation. After 5 hours of manipulation in the catheterization laboratory, the procedure was aborted because the MPA stent migrated to the RV. The patient was urgently transported to the main OR, and an open-heart surgery was performed to retrieve the migrated stent. Pulmonary valve replacement and pericardial patchplasty of the MPA and RV outflow tract were also performed. Later, the patient was discharged home on POD #10.”

7. Images: "Echo and/or Fluoroscopy of displaced stent will be presented.

8. Discussion:

Patients with TOF enjoy a high survival rate following an early surgical repair. However, as these patients age, the sequelae of the initial correction requires repetitive interventions for pulmonic stenosis and/or pulmonic insufficiency. Recent advancements in the percutaneously placed valves have led to a decrease in secondary open-heart surgeries and to an increase in minimally invasive catheterization techniques.

Unfortunately, the reported rate of transcatheter valve placement failure requiring conversion to an open-heart surgery after pulmonary outflow manipulation can be more than 3% (4). Another retrospective study demonstrated that 21% of children and young adults with different congenital heart diseases needed urgent surgery after stent implantation (5).

The trend of increasing TOF patient population with increase in performance of percutaneous valve techniques, suggests that anesthesia providers will witness a greater number of conversions of minimally invasive procedures to open-heart surgeries during their career.

Our case demonstrates the importance of having a promptly available Cardiac surgeon, Cardiac OR, and Congenital Cardiac Anesthesiologist when having a patient scheduled for minimally invasive PA intervention in the Catheterization laboratory.

9. Detailed case description:

A 16-year-old Male with PMH of von Willebrand's disease (vWD) and VACTERL association congenital defects (TOF, bilateral clubfeet, imperforate anus, gut malformation, tethered spinal cord), s/p TOF repair with a large transannular RVOT patch at 7 months of age. He required bilateral percutaneous pulmonary branch balloon dilation arterioplasties with the placement of LPA stent at 2 years of age, and a replacement of a right ventricle to pulmonary artery homograft conduit with bilateral branch pulmonary patch angioplasties and postoperative placement of RPA stent at 2.5 years of age. At 3 years of age, he underwent percutaneous balloon dilation of prior RPA stent and reinsertion of LPA stent with balloon dilation angioplasty.
An echocardiogram performed prior to this scheduled procedure showed a good homograft function with mild stenosis (8/16 mmHg) and moderate pulmonic insufficiency, moderate dilatation of RV with normal systolic function, and LVEF of 53% by 2D measurement. The patient was asymptomatic without decrease in exercise tolerance, METS >7.

A PA stent re-exploration and a Melody TPV implantation was planned to address pulmonic insufficiency and to preserve RV function. He received one dose of Humate P preoperatively for his vWD. Catheterization laboratory procedure was performed under General anesthesia with ETT placement. Angiography revealed mild calcification of the RV-PA conduit with a minimum diameter of 18mm, RV heavily trabeculated and moderately dilated, RV with low normal systolic function. The RPA stent was found to be protruding into the MPA. The interventional plan involved stenting of the MPA and Melody valve implant. After 5 hours of manipulation, the procedure was aborted because the MPA stent migrated to the RV. Right after this, the patient exhibited frequent ventricle ectopy without blood pressure compromise. The patient was urgently transported to the main OR for open-heart surgery. Retrieval of the migrated stent, pulmonary valve replacement (23 mm Carpentier-Edwards), and pericardial patchplasty of MPA and RV outflow tract were performed. Before the initiation of cardiopulmonary bypass, the patient had two runs of V-fib which were electro converted. Bypass time was 100 min, and Humate P was re-dosed after bypass.

The postoperative course was complicated by mild chest effusion. The patient was discharged home on POD #10 but had to be readmitted in 3 days due to pleural effusion with respiratory compromise.

10. Literature:


