Percutaneous transvenous mitral commissurotomy in a case of situs inversus, dextrocardia and rheumatic mitral stenosis

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ABSTRACT: Situs inversus with dextrocardia is a rare congenital disorder. There is rare coincidence of rheumatic severe mitral stenosis in a patient with situs inversus and dextrocardia. Technical difficulties for doing percutaneous transvenous mitral commissurotomy (PTMC) in such patients are well known and there are few reports of PTMC in situs inversus with dextrocardia. Here we report a case of 53 year old female with situs inversus and dextrocardia where PTMC was successfully done with a few modifications of standard Inoue technique. The patient had dyspnea on exertion of NYHA class III with initial mitral valve area of 0.8 cm² and severe pulmonary arterial hypertension with PA systolic pressure of 106 mmHg. Femoral vein access was performed from the left side to align the septal puncture needle and balloon to the left sided IVC and to facilitate LV entry. Septal descent of the septal puncture needle (Brockenbrough needle) was performed in the AP view with the needle rotated from 12 o’clock to 7 o’clock position instead of 5 o’clock position. The right border of the spine was used as a landmark for trans-septal puncture. The LA pressure fell from 19 mmHg to 9 mmHg with no residual gradient across the valve. PA systolic pressure dropped to 48 mmHg. Echocardiography showed a well-divided anterior commissure with a MVA of 1.8 cm² and mild mitral regurgitation with mild pulmonary hypertension. In summary, PTMC is feasible in the rare patient with dextrocardia with additional modifications of the Inoue technique and protocols for groin and septal puncture.

KEY WORDS: Mitral commissurotomy; Inoue technique; Situs inversus; Dextrocardia; Rheumatic mitral stenosis

INTRODUCTION

Variation in cardiac anatomy results in technical difficulties during percutaneous transvenous mitral commissurotomy (PTMC). There are only few reports on successful PTMC in abnormal cardiac anatomy using the standard Inoue technique. Here we describe a case of a 53-year-old female with situs inversus and dextrocardia, where PTMC was successfully performed with a few modifications of the standard Inoue technique.

CASE DETAILS

A 53-year-old female with a history of dyspnea on exertion of NYHA class III of 2 months’ duration.

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Clinical, radiological (Fig 1) & echocardiographic evaluation suggested situs inversus, dextrocardia and severe rheumatic MS. Her mitral valve area (MVA) was 0.8 cm² with pliable valve and moderate subvalvular pathology. The gradient across the mitral valve had a peak of 28 and mean of 17 mmHg. There was severe functional tricuspid regurgitation with pulmonary artery systolic pressure of 106 mmHg. PTMC was planned. Femoral arterial and venous punctures were made on the right side for LV and PA catheterization (Fig 2). Right heart catheterization was performed and it showed severe pulmonary arterial hypertension. Left femoral vein access was done for septal puncture (Fig 2). Septal descent of septal puncture needle (Brockenbrough needle) was performed in the AP view with the needle rotated from 12 o’clock to 7 o’clock position instead of 5 o’clock position. The right border of the spine was used as a landmark for trans-septal puncture with a
site midway between pigtail catheter and spine (Fig 3 and 4). The IAS was dilated with a septal dilator, a 26 mm Accura balloon was tracked over the wire to the LA and LV entry was done (Fig 5). Inflation was done at 22 mm. The LA pressure fell from 19 mmHg to 9 mmHg with no residual gradient across valve. PA systolic pressure was dropped to 48 mmHg. Echocardiography showed a well-divided anterior commissure with a MVA of 1.8 cm² and mild mitral regurgitation with mild pulmonary hypertension. At 6 months follow up patient remained in class I.
DISCUSSION

Dextrocardia has an incidence of 1:10,000 and is a rarer cardiac anomaly when occurring along with severe rheumatic MS. Here dextrocardia is a coincidental occurrence. There are a few case reports in the literature on PTMC in similar settings due to the fact that many of these patients undergo surgical commissurotomy due to the technical difficulties involved. In general, anatomical variations of the heart are considered relative contraindications for transseptal catheterization, as it is thought to have a higher risk of cardiac perforation.

Transseptal catheterization was performed from the left groin to align the tracking of balloon and septal puncture needle with IVC and common iliac vein and facilitate LV entry. The delineation of the IAS is important and difficult step in the procedure. Verma et al used levo phase pulmonary angiography for IAS delineation in a patient with isolated dextrocardia and normal atrial situs. This step had not thus far been described in any patient with mirror-image dextrocardia undergoing PTMC. The catheter placed in the non-coronary aortic sinus can mark the antero-superior limit of the IAS also as in this case.

The radiographic images acquired in the inverted position can be used as fluoroscopic guidance for the septal puncture, as previously described by Nallet et al. The inverted fluoroscopic settings, which simulates normal anatomy, facilitates manipulation of the transseptal needle and balloon in the LA. These modifications supplemented by contrast injection into the septum to delineate the IAS, can be of additional use in the presence of the septal aneurysm. The usefulness of intracardiac echocardiography (ICE) in preventing serious complications in transseptal procedures when the cardiac anatomy is unusual or distorted has also been highlighted recently.

Safety and efficacy of this type of procedure has been documented in the past. The protocol for groin puncture, septal puncture and LV entry which are used in this case have been tried and regarded as safe in the past by demonstration in patients with sites inversus and dextrocardia. The use of intraoperative trans-esophageal echocardiography may also be helpful when needed. The followings are the sites of groin puncture for balloon in different cardiac mal-positions:

- Situs solitus and levocardia – Right
- Situs solitus and dextrocardia – Right
- Situs inversus and levocardia – Left
- Situs inversus and dextrocardia - Left

In summary, PTMC is feasible in the rare patient with dextrocardia with additional modifications of the Inoue technique and a few protocols for groin and septal puncture.

REFERENCES