pathophysiological mechanisms are highly variable. Among patients undergoing coronary angiography, the incidence depends on the population studied and the criteria used to define an anomaly. One large study reported an incidence rate of coronary anomalies of 1.3% for adults undergoing catheterization primarily for the detection of coronary artery disease. This is the first ever study conducted in western Rajasthan to detect the incidence and pattern of coronary anomalies.

Methods: We retrospectively studied 8500 coronary angiographies (CAG) done in our institution over a period of 12 years from 2004 to 2015 for detection of incidence and pattern of coronary anomalies. Patients with ischemic heart disease and valvular heart disease who underwent CAG were included in the study. Patients with congenital heart diseases were excluded from the study. Patients with other coronary anomalies like ectasia, myocardial bridging, abnormal high and low origin of coronary arteries from normal sinus and separate origin of the conus artery from the right coronary sinus (RCS) were also excluded from the study.

Results: Out of the 8500 angiograms screened, a total of 108 coronary anomalies were detected (incidence of 1.27%). Anomalies of origin and course was the most common anomaly (106 out of 108 patients) followed by anomalies of coronary termination (fistulas) which was seen in just two patients. Most common anomaly was absence of left main artery with separate origin of the left anterior descending (LAD) artery and left circumflex artery (LCx) ($n = 36$, 33.3%), followed closely by anomalous origin of right coronary artery (RCA) from left sinus ($n = 34$, 31.48%). Anomalous origin of LCx from right sinus/RCA was the third most common anomaly ($n = 22$, 20.37%). Other rare anomalies include anomalous origin of left coronary artery from right coronary sinus ($n = 6$, 5.55%), RCA from posterior sinus ($n = 4$, 3.7%). Single coronary artery, LAD from RCA and coronary artery fistula were seen in two patients each ($n = 2$, 1.85%)

Coronary anomaly	Number of patients ($n = 108$)	Angiographic incidence %	Anomaly incidence %
Separate origin of LAD and LCX	36	0.42	33.3
RCA arising from LCS	34	0.40	31.48
LCX arising from RCS/RCA	22	0.25	20.37
LCA arising from RCS	6	0.07	5.55
RCA arising from posterior sinus	4	0.04	3.70
Single coronary artery	2	0.02	1.85
LAD from RCA	2	0.02	1.85
Coronary artery fistula	2	0.02	1.85

Conclusion: In our study though the total incidence of coronary anomalies was similar to that in other studies, the pattern of coronary anomalies was slightly different from that reported from different parts of the world.

Beauty of device closure in RSOV

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Introduction: Sinus of valsalva aneurysm is a rare congenital abnormality. When it ruptures, mostly to the right heart resulting in left-to-right shunt, the patients may experience severe heart failure. We here present a case of ruptured sinus of valsalva (RSOV) to right atrium which was closed by PDA device. Patient despite all odds finally survived and now leading a normal life.

Case report: A 40-yr-old female patient presented to us with dyspnea and palpitations since 1 week. On examination, there was bounding pulse with elevated jugular venous pulse (JVP). Examination revealed continuous murmur in lower sternal border and ECG showed atrial fibrillation. Further, 2D Echo done showed aortic sinus aneurysm arising from non-coronary cusp, which had ruptured into the right atrium. Patient was diagnosed to have RSOV to right atrium and was given options of device closure or surgery. As patient had some financial problem, patient asked for some time to make necessary arrangements and then got discharged. Patient was discharged with diuretics, beta blockers.

Patient did not turn up for 2 months and later when she visited us after 2 months she was extremely cachexic. She had severe nausea, vomiting, weight loss of about 15 kg. On examination, she had elevated JVP (15 cm of water) and gross congestive hepatomegaly (7 cm from costal margin) and was in atrial fibrillation. Looking at her condition this time surgeons also refused surgery. And then we planned for device closure.

The procedure was performed under local anesthesia. A PDA device (LIFE TECH) of size 14/12 mm was chosen to close the defect. Patient tolerated the procedure very well and 2D Echo done showed no AR, TR or residual shunt. Patient was put on prophylactic antibiotics and on aspirin 75 mg. Patient later started taking feeds and her appetite improved.

But this was not the end of story. On 3rd day after procedure, patient developed recurrent episodes of VT and continuous marathon CPR and DC was given. At one stage we thought stopping our resuscitation measures, but luckily by that time our ABG report came which showed hypokalemia and hypocalcaemia, which was the culprit. Which might have occurred due to refeeding syndrome (this syndrome has been described in extremely cachexic patients in whom refeeding leads to electrolyte disturbances). Kcl and calcium gluconate infusion with inotropes was given with careful monitoring. And by god's grace she later gradually improved and her pre discharge echo showed 2D LVEF 15–20% and she was discharged on 12th day. Now, she is coming for regular follow-up and her symptoms have improved from NYHA 4 to NYHA 1 and she has regained her weight and now in normal sinus rhythm. 2D Echo showed no residual shunt, decreased PA pressures, and 2D LVEF 50–55%.

Implication to clinical practice: For patients with RSOV, although conventional surgical correction under cardiopulmonary bypass carries low mortality, postoperative septicemia, infective endocarditis, and prolonged recovery time make percutaneous device closure an attractive alternative. This case clearly demonstrates the beauty of device closure.

A rare association – Noonans syndrome with coarctation of aorta and cleft anterior mitral leaflet

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Introduction: Noonans syndrome is a multisystem disorder, autosomal dominant with variable penetrance, and with an estimated prevalence of 1 in 1000 to 1 in 2500 live births. It is characterized by distinctive facial features, short stature, chest deformity, and congenital heart disease. It is diagnosed clinically but genetic mutation can be identified in 61% of cases. In 1962, Jacqueline Noonan, a pediatric cardiologist described 9 cases whose faces were remarkably similar. The physical findings are short stature,

wide spaced eyes, prominent epicanthal folds, ptosis, ears are low set, webbing of neck, pectus deformity of chest, and wide spaced nipples. The mutation in the Ras/MAPK pathway – PTPN11 gene, KRAS, SOS1, RAF1, SHOC2 gene mutation isolated in Noonans syndrome. Cardiovascular involvement is seen in 80% of cases, pulmonary valve stenosis most common, in 25–35% cases valve is dysplastic and associated with ASD, hypertrophic cardiomyopathy is present in 20% of cases with variable presentation and severity. Left sided obstructive lesions may develop in adulthood, coarctation of aorta occasionally described with 12.8% of cases, mitral valve anomalies have been described.

Case report: A 12-year-old child presented with class-2 breathlessness, poor gain in weight on examination, patient pulse rate = 78/mt with radio-femoral delay, blood pressure in upper limb – 130/90 and lower limb was 80/60 mm of Hg, epicanthal folds, hypertelorism, low hair line, webbed neck cvs-s1s2, ejection click with ESM, RS-normal, chest X-ray was showing rib erosion and 2D echo was showing gradient across the aorta with AML cleft mitral valve with severe eccentric mitral regurgitation. Aortic angiography was done, coarctation confirmed with gradient of 58 mm of Hg. Patient was referred to CT SURGERY department for surgical management.

Discussion: Coarctation of aorta accounts 6–8% of congenital heart disease. This can occur in isolation or associated anomalies, bicuspid aortic valve (85%), parachute mitral valve, shunt lesions like patent ductus arteriosus, ventricular septal defect, cerebral aneurysm (3%). Coarctation of aorta symptomatic in early infancy and after 20–30 years. The major symptoms are congestive heart failure, rupture or dissection of aorta, infective endocarditis, cerebral hemorrhage. When associated with mitral regurgitation will increase left ventricular end diastolic volume and preload. This can be associated with syndromes like Turners syndrome, Noonans syndrome. Usually chest X-ray may show notching of ribs, 2D echo may show gradient across the aorta by Doppler evaluation, angiography remains the gold standard evaluating coarctation of aorta. It can be treated surgically or by percutaneous intervention. Surgically, coarctation treated with resection and end-to-end anastomosis, prosthetic patch aortaplasty, left subclavian flap aortaplasty. Percutaneous balloon angioplasty and stenting as percutaneous intervention.

Conclusion: A rare association – Noonans syndrome with coarctation of aorta and cleft anterior mitral leaflet is being reported.

Lutembacher syndrome

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Lutembacher's syndrome is a rare heart disease, comprises of ASD (congenital or acquired) ostium secundum defect with mitral stenosis (rheumatic origin). The patient Mrs. Gangamma a 50-year-old woman, nondiabetic, nonhypertensive, nonsmoker admitted in our hospital (RRMCH Bangalore) on 22/01/2015 with the complaints of breathlessness and chest pain for 4–5 months, cough for 4–5 months. Breathlessness is more in lying position and also during exertion. Her breathlessness status is qualified as NYHA–class-III (New York Heart Association). Patient is a known case of Rheumatoid Arthritis from past 10 years. There was history of recurrent attack of Rheumatic fever. And within last 2 years, she developed several episodes of respiratory distress. Subsequently, she developed MS from rheumatic carditis. ASD was congenital in origin. On examination, pulse was 80 beats/min, small volume pulse, regular, BP – 120/80 mmHg, JVP was raised, ankle odema present. Precordial examination reveals early systolic murmur on pulmonary area and mid diastolic murmur on mitral area. Respiratory system examination reveals bilateral basal creps. If diagnosis could be done earlier, surgical catheter closure of ASD with replacement of mitral valve bears a good prognostic value. Our patient has moderate MS (MVOA: 1.2 cm^2) with severe pulmonary hypertension (PASP: 65–70 mmHg). So early PTMC (percutaneous transmitral commissurotomy) of the MS gives good clinical improvement for the patient. Our patient was treated with digoxin, diuretics, ACE inhibitors and the patient improved significantly. So here is a rare case of Lutembacher's syndrome (moderate MS with ASD ostium secundum defect with severe PAH) associated with rheumatoid arthritis.

Palliative catheter intervention in adult tetralogy of Fallots

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Introduction: Tetralogy of Fallot is the most common cyanotic congenital heart disease reaching adult life without surgical intervention. Surgical intervention in adult Tetralogy of Fallot is challenging and often complicated by residual cardiac defect.

Methods

1. Of 38 patients with diagnosis of adult TOF (between Jan 2012 and Jan2014) 5 required palliative catheter intervention procedures.
2. Patients (deemed high risk for intracardiac repair) with prior history of BT shunt underwent stent angioplasty to BT shunt with excellent results. Both patients underwent intracardiac repair later.
3. Patient (aged 23 years) presented with a resistant hypercynotic spell due to hyperviscosity syndrome. He underwent palliative RVOT stenting with excellent result. He underwent intracardiac repair with RVOT reconstruction with bioprosthetic valve with excellent result.
4. Patient (age 17 years) after intracardiac repair had a very large VSD. He was ventilator and inotrope dependent for a week. He was subjected to VSD device closure with excellent outcome.
5. Patient developed severe pulmonary artery bifurcation stenosis after RV-PA conduit repair. He underwent bilateral PA angioplasty with balloon expandable stents (SKS strategy) with excellent outcome.

Result: All 5 patients with palliative catheter intervention had an excellent outcome. They are on close medical follow up.

Conclusion: Palliative catheter intervention in patients with adult TOF plays a significant role in improving overall outcome in this challenging subset of congenital heart disease.