suggestive of pulmonary venous obstruction. This was confirmed on transoesophageal echocardiography. Patient was posted for percutaneous balloon angioplasty of pulmonary veins. After right femoral venous puncture, the wire did not cross the IVC. Hence check angiography was done through right femoral venous sheath which showed total occlusion of IVC in lower abdomen with reformation of IVC in the upper abdomen through tortuous collaterals arising from left common iliac vein. There was no channel through which the wire could reach the RA safely. Hence the procedure was abandoned and the patient was posted for surgical correction of pulmonary vein stenosis. After the surgical reconstruction, patient developed sudden right heart failure and subsequent cardiogenic shock, renal failure, and sepsis. The patient was intubated and kept on mechanical ventilator, managed with ionotropic support, intravenous antibiotics and three cycles of hemodialysis. The patient was discharged after 10 days of surgical correction.

The pulmonary hypertension crisis at the time of presentation led to hypoxemia that triggered an increase in pulmonary vascular resistance, increased hydrostatic pressure, and worsened pulmonary edema. Given the extent of the baseline pulmonary hypertension in our patient, pulmonary vascular recruitment was likely limited, which explained the inability of the pulmonary circulation to accommodate an acute increase in pulmonary pressures. The latter, in combination with the underlying broncho-pulmonary dysplasia, may account for the patient's cardiorespiratory failure despite unilateral disease. In the small group of patients with diffusely hypoplastic pulmonary veins, the term ''primary'' pulmonary vein stenosis is preferred. The reason for this difference in terminology is that it is becoming more apparent that the disease is often progressive and may not even be evident at birth.

Percutaneous balloon valvuloplasty with inoue balloon catheter technique for pulmonary valve stenosis in adolescents and adults

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Background: Percutaneous balloon valvuloplasty (PBV) is the procedure of choice for uncomplicated severe or symptomatic pulmonary stenosis. Current approaches utilize various fixed size balloon catheters using a single or double balloon technique. The present report describes our experience in BPV using the inoue balloon catheter in adolescent and adult patients.

Aim: To assess the immediate and mid-term outcomes of PBV for pulmonary stenosis with a single inoue balloon catheter in adolescent and adult patients.

Methods and results: Between June 2010 and July 2015, we performed percutaneous pulmonic valvuloplasty with a single inoue balloon catheter in 32 adolescent or adult patients (19 females and 13 males) aged 8–54 years (23.6 ± 11.5). Majority of patients presented with exertional dyspnea (87.5%) while 3 patients had pedal edema and 2 complained of syncope. Pulmonary annulus on catheterization was (18.8 ± 2.2 mm) and balloon-pulmonary annulus ratio was (1.31 ± 0.16). The mean right ventricular systolic pressure and the pulmonary valvular peak-to-peak systolic gradient decreased from (121.6 ± 42.4 to 61.19 ± 24.5 mmHg) and (100.9 ± 43.3 to 36.4 ± 22.5 mmHg), respectively, following PBV. Post procedure, reactive right ventricular outflow tract stenosis was seen in 5 patients and mild pulmonary regurgitation was detected in 10 patients. Clinical and Doppler echocardiographic follow-up studies were performed 0.2–5 years after the procedure in 17 patients. All patients were asymptomatic at follow up. Right ventricular outflow tract stenosis was attenuated in all 5 patients on follow-up. There was no increase in grade of pulmonary regurgitation. There was no restenosis in the followed-up patients with maintenance of good RV function [RV-PA gradient and TAPSE on echocardiography (19.94 ± 8.46 mmHg) and (22.7 ± 1.49), respectively].

Conclusions: Patients with congenital pulmonic stenosis who present in adolescence or adult life can be treated with PBV using inoue technique with excellent short- and intermediate-term results.

Congenital heart disease: Spectrum and distribution at a tertiary health care centre in western India

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Background: Changing pattern and incidence of congenital heart diseases (CHD) have been observed in various geographical locations.

Aim: To study the frequency, age-wise distribution, and spectrum of CHD at a tertiary health care centre in Ajmer, Rajasthan.

Methods: A retrospective analysis of case records of 8641 patients in the age group of 0–18 years from January 2008 to July 2015 was done to ascertain the spectrum and distribution of CHDs. Clinical examination, electrocardiography, chest X ray, and transthoracic echocardiography were used as diagnostic tools.

Results: Out of 8641 patients, 2052 (23.75%) were found to have CHD. Male preponderance was observed (male to female ratio = 1.43). Study group comprised of 12.62% neonates, 39.38% infants, and 47.81% of more than 1-year age. A total of 1742 (84.89%) were acyanotics, and 310 (15.11%) suffered from cyanotic heart disease. Among the acyanotic heart diseases, ventricular septal defect (VSD) was the most frequent lesion seen in 700 (40.18%), followed by atrial septal defect (ASD) in 370 (21.24%) children. Among the cyanotic heart diseases, tetralogy of Fallot (TOF) was the most frequent cyanotic heart disease seen in 196 (63.23%) patients.

Conclusion: The frequency of CHD at a tertiary care centre in western India was 23.75%. VSD and ASD were the most common acyanotic while TOF was the commonest cyanotic congenital heart defect observed. TTE plays a major role in the diagnosis of CHD. When clinical evidences lead to suspicion of congenital heart defect, an echocardiography should be performed.

Overall distribution			
Acyanotic (n = 1742, 84.89%)		Cyanotic (n = 310, 15.11%)	
VSD	700 (40.18%)	TOF	196 (63.23%)
ASD	370 (21.24%)	TGA	32 (10.32%)
PDA	163 (9.36%)	DORV	18 (5.81%)
PS	134 (7.69%)	AVSD	20 (6.45%)
AS	33 (1.89%)	Single ventricle	10 (3.22%)
Others	342 (19.64%)	Others	34 (10.44%)

Neonates (up to 1 month)			
Acyanotic (n = 233, 89.96%)		Cyanotic (n = 26, 10.04%)	
VSD	76 (27.47%)	TGA	12 (46.15%)
ASD	64 (32.62%)	TOF	4 (15.38%)
Infants (1 month to 1 year)			
Acyanotic (n = 684, 84.65%)		Cyanotic (n = 124, 15.35%)	
VSD	318 (46.49%)	TOF	72 (58.06%)
ASD	146 (21.35%)	TGA	12 (9.68%)
Above 1year			
Acyanotic (n = 825, 83.69%)		Cyanotic (n = 160, 16.31%)	
VSD	306 (37.27%)	TOF	120 (75.0%)
ASD	160 (21.35%)	TGA	8 (5.0%)
PDA	92 (11.21%)	DORV	12 (7.5%)

VSD device closure with PDA device

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Background: Transcatheter closure of ventricular septal defect using PDA devices is gaining acceptance in selected cases as compared to surgical closure which is associated with morbidity and mortality. Potential advantages of the transcatheter closure over conventional surgery include a smaller incision, shorter stay, and fewer complications.
Methodology: A total 78 cases underwent echo and cardiac cath. Out of 78 patients, 10 patients sent to surgery because of improper size or location of the VSD. A total of 68 cases were having defect size ranging from 3 mm to 10 mm. The location of VSD was perimembranous in 60 cases, muscular in 08 cases, and apical in 01 case. The procedure was performed in 68 cases with PDA device.
Procedure: LV angiography showed VSD. LIMA catheter was passed in LV with rotation and pull-back technique, then crossed VSD and was placed in RV. Double length Terumo 0.035″ × 260 cm wire was taken and passed through RV, RA, IVC, SVC, or PA. Snared through femoral venous and arterio venous loop was created with the help of Terumo wire. PDA sheath was passed through RA, RV and then into aorta with Kissing technique sheath was positioned in LV. PDA device was loaded in sheath. Device was positioned on LV side first and then on RV side. Position was checked on 2D-Echo which was found to be accurate. Wire and catheter was removed. Post procedure treatment with steroids was advised as per standard treatment. Anti-platelet aspirin was recommended for one month.
Results: Procedure was done successfully in 66 without any complications. In 15 cases, transient BBB was developed on during procedure. In 1 case, complication of CHB occurred during procedure in which temporary pacemaker (TPM) was implanted for 3 days. In another case, complication of CHB occurred in ICU after 3 h in which TPM was done for 2 days. In 2 cases, device was embolized, 1 device successfully retrieved with help of the basket snare and continued procedure with bigger size device, another 1 case went to surgery. A total of 1 case of death was observed on 5th day, which was due to stroke. At 1-year follow-up, no shunt was noted in ECHO and ECG remained same with no additional conditional defect.
Conclusion: Transcatheter closure is safe and efficacious in selected cases of perimembranous and muscular VSD using PDA device.

Syndromic tetralogy of Fallot – A case report

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Introduction: Tetralogy of Fallot (TOF) is the most common cause of cyanotic congenital heart disease in ventricular septal defect and pulmonary stenosis physiology. TOF is associated with many syndromes, the most common being Down syndrome, Velocardiofacial syndrome, Holt Oram syndrome, and Poland's syndrome. Incidence of VACTERL syndrome is rare compared to the above mentioned syndromes.
Case report: A 12-year-old boy was brought with complaints of shortness of breath and palpitations on exertion. He had history of squatting episodes. Prenatal and perinatal history in mother was normal. He attained developmental milestones on par with his peers. His weight was <2SD for his age, mid arm circumference – 13 cm. On examination he had dysmorphic features – left absent thumb, kyphoscoliosis, low set ears, low hair line, hypoplastic mandible, deciduous teeth, radial hypoplasia with pandigital clubbing, and JVP was normal in height and waveform. Cardiovascular examination – LV apex, parasternal heave 3/3, systolic thrill left 2,3 ICS.S1 normal, S2 normal split, Grade 5/6 MSM at left 2nd and 3rd ICS selective conduction to left side. He was evaluated. Hb 13.5 gm/dl, WBC counts were within normal limits. The following are noted, ECG – normal sinus rhythm; extreme right axis deviation −150°; RVH with strain; Katz Wachtel phenomenon; Chest X ray – oligemic lung fields; and no cardiomegaly. ECHO showed situs solitus, large subaortic ventricular septal defect with overriding of aorta <50%, with severe valvular pulmonic stenosis and subvalvular infundibular dynamic obstruction. The gradients across the pulmonic valve were 92/63 mmHg and at the infundibulum was 53/14 mmHg. Pulmonic valve was doming. RVH was present with free wall thickness of 0.7 cm. Based on the dysmorphic features, he was categorized as syndromic TOF.
Discussion: VACTERL is an association of nonrandom occurrence of Vertebral, Anorectal anomalies, Cardiac anomalies, Tracheoesophageal anomalies, Radial hypoplasia/renal anomalies, and limb defects. All features need not present at a single time for diagnosis. According to Genetic Home Reference and National Library of Medicine (ghr.nlm), three anomalies ought to be present for labelling a patient to have VACTERL association. The present patient had Vertebral anomalies (Scoliosis), Cardiac anomalies (TOF), Renal anomalies (left kidney shrunken), Radial hypoplasia, absent thumb. The close differential diagnosis for this patient was Holt Oram Syndrome, it can be excluded in individuals with congenital malformations involving – ulnar ray only, kidney, vertebra, craniofacies, auditory system (hearing loss or ear malformations), lower limb, anus, or eye.
Clinical implications: TOF association with VACTERL is rare and hence reported.

A study of incidence and pattern of coronary artery anomalies in western Rajasthan

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Background and objectives: Coronary artery anomalies are a diverse group of congenital disorders whose manifestations and