alone or improvement in PA saturations alone. Likewise a fall in PAEDP and poor improvement in saturations leads to higher post-operative residual PAH and longer ICU ventilator need and need for milrinone to counter postoperative residual PAH.

Comparison of ambrisentan with sildenafil in congenital heart disease with irreversible pulmonary artery hypertension

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Background: The pathogenesis of PAH involves endothelial dysfunction characterised by over-expression of vasoconstrictors such as endothelin-1 (ET-1) and thromboxane A2. Endothelin receptor antagonists (ERAs) are a class of potent vasodilators and antiinmitotic substances. Aims & Objectives - To assess the efficacy of endothelin receptor antagonists (ERAs) Ambrisentan in the treatment of patients with PAH secondary to congenital heart disease.

Methods: It is a retrospective observational study, carried out at single tertiary care center. Patient of acyanotic congenital heart disease (e.g.VSD, PDA, AV canal defect) admitted in cardiology department, found to have irreversible pulmonary hypertension on cardiac catheterization were enrolled in study. A total of 42 patients were enrolled in the study, 24 patients had received 10 mg of ambrisentan and remaining group 20 patient had received sildenafil at maximum tolerated dose for at least 6 months.

Primary outcomes were exercise capacity (a six-minute walk test (6MWD)) and World Health Organization (WHO) functional class. Secondary outcome- Adverse events (for example, hepatic toxicity).

Results: An increase in the 6MWD was reported in the ambrisentan group. Increases in the 6MWD at 24 weeks was 51 meters in Ambrisentan group compared to 20 meters in sildenafil group.(95% CI 27–76; P, 0.001). Patient enrolled in the study were prominently WHO functional classification II (29%) and III (65%). Study demonstrated a significant improvement in WHO functional classification for patients receiving ambrisentan as compared with sildenafil (P = 0.036). Of the patients receiving ambrisentan, 2 (9.7%) discontinued because of adverse events (gastroenteritis, headache, face edema [n=1 for each]). Side effect of ambrisentan was found to be headache 2 (9.0%), peripheral edema 4 (18.18%). No evidence of hepatotoxicity like elevated liver enzymes, bilirubin in ambrisentan group.

Conclusion: Study demonstrate the efficacy and safety of ambrisentan in the treatment of patients with symptomatic PAH. The favorable efficacy-to-safety profile of ambrisentan may offer potential advantages over the currently approved treatment options.

Our experience of percutaneous ventricular septal device closure at our centre (tertiary care) – Retrospective study

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Background: Ventricular septal defect (VSD) is the most common congenital heart defect at birth, as well as one of the common heart defects in adult age group. Repair of ventricular septal defect (VSD) has been historically performed surgically. However, percutaneous VSD closure is also feasible and the frequency has increased given the desire of young patients to avoid surgery. Standard treatment of VSD is open surgery, which is widely performed with but still carries risks, such as complete atrioventricular block, residual shunt, postpericardiotomy syndrome, and wound infection. Transcatheter VSD device closure is a treatment option for isolated uncomplicated muscular VSDs, and for membranous VSDs, in selected patients with suitable anatomy. Appropriate anatomy for transcatheter closure includes a VSD location remote from the tricuspid and aortic valves with an adequate rim of tissue.

Here we are presenting our experience about percutaneous VSD device closure or attempted VSD device closure in 12 patients at tertiary care centre.

Methods: Between June 2013 and July 2014 patients undergoing percutaneous VSD device closure or attempted VSD device closure were studied retrospectively. All patients underwent detailed conventional two dimensional and colour Doppler transthoracic echocardiography to study the type of VSD, shunt across the VSD, length of aortic rim and pulmonary hypertension. VSD size was confirmed angiographically and device closure was done under general anesthesia with TEE guidance according to standard protocol. During immediate postoperative period, rhythm and position was monitored by doing ECG and TTE respectively. Patients were given weight based aspirin and clopidogrel for 6 months. Patients were followed up on outpatient basis every 6 months for 1 yr and then every year and TTE was done during each visit.

Result: There were total 12 patients out of which successful device closure was done in 10 patients. The average age of patient was 6.9 yrs (ranges 2.5 to 18 yrs). The 5(41.6%) patients were female and 7 (58.3%) were male. The median size of VSD was 5.3 mm (ranges 2.3 to 16 mm) one (8.3%) patient had two VSD. Average size of device used was 7.1 mm .procedure related complication occurred in 4 patients. One (8.3%) patient had CHB after releasing of LV disc hence procedure was abandoned. One (8.3%) patient had embolization of device in the aorta which was successfully retrieved by transcutaneous route. No patient had postprocedure death. No Patients had postoperative rhythm disturbances. All patients were discharged two days after procedure. All patients were follow up at six month and 1 year by ECG and transthoracic echocardiography.

Conclusion: In experienced hands, transcatheter VSD closure can be performed safely and successfully with low morbidity and mortality. Transcatheter approach provides a less-invasive alternative that may become the first choice in selected VSD patients.

Experience with large ostium secundum atrial septal defect (ASD) closure by transcatheter percutaneous device (device) at J.J.Hospital

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Background: Small ASD are routinely closed by device. Larger ASD by device closure is problematic. But, in these defects also, device closure has high success. We present our experience of device closure of ASD measuring >25 mm.
Methods: We evaluated 2 years records of device closure. ASD >25 mm attempted for device closure were studied. They underwent transthoracic 2D Echocardiography (TTE), TEE to characterize defect. Size was determined angiographically/balloon sizing during procedure. Device was made up of Nitinol wires filled with polypropylene or polyester. Device was placed across ASD with/without balloon support. Defect in which, device could not be positioned even after balloon support, was deferred. ASD device was released under TEE to confirm proper position. Postoperatively, rhythm and position was monitored by ECG and TTE respectively. Those having displacement of the device were shifted for surgery. Patient was given aspirin and clopidogrel postop. Patients were followed up on OPD basis and TTE was done during each visit.

Results: 26 patients were attempted for device closure. 12 males and 14 females. Mean age-16.76 years. Mean ASD size-30.67 mm (range-25.2-36.2). Mean IAS size-45.36 mm. Mean size of rims-7.5 mm atrioventricular rim, 7.6 mm interatrial rim, 4.5 mm aortic rim, 5.5 mm of IVC rim and 6.3 mm of SVC rim. Mean device size-32.33 mm. 2(7.69%) defects could not be closed because of inability to position the device. 2(8.83%) devices displaced within mean 8 hours and patients were shifted for surgery.

During immediate postoperative period, 2(8.33%) had junctional rhythm and 1(3.84%) had AV dissociation. All disturbances normalized in Mean 27.6 hrs. Mean hospital stay-86 hrs with de- rhytym and 1(3.84%) had AV dissociation. All disturbances

Complex TAPVC — Challenges and outcomes in our institute

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Background: Management of Total anomalous pulmonary venous connection becomes complex when it is associated with other intracardiac anomalies. Perioperative mortality and morbidity increases significantly. We present our modest experience with the management of complex TAPVC in our hospital.

Methods: Our study group includes eight patients with TAPVC associated with other intracardiac anomalies. There were 3 girls and 5 boys. Four of them were less than 5 kg in weight, and the other 5 were more than 5 kg in weight. Four patients had severe PAH and 4 patients had pulmonary stenosis. Five patients were of supracardiac type and one was cardiac type. Two patients had DORV, Two had TOF, one had dTGA, one had Truncus Arteriosus, two patients had atrioventricular canal defect. 2 patients had pulmonary vein obstruction.

Results: All patients underwent rerouting of pulmonary veins. Concomitant procedures included intraventricular tunnel repair of VSD and infundibular resection in DORV patient. Two patch technique AV canal repair was done for AVSD patient. SVC plasty, atrioventricular canal repair and PA banding were done in unbalanced atrioventricular septal defect with DORV. Intracardiac repair through transtial approach was done for tetralogy of Fallot. Right ventriculopulmonary artery conduit was done for truncus arteriosus. Arterial switch was done for TGA patient. There were 2 hospital deaths.

Conclusion: Precise surgical techniques and attentive perioperative care will reduce the mortality and improve the outcome though the management of Complex TAPVC is technically challenging.

Evaluation of anomalous coronary origin and course on different imaging modalities

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Introduction: The coronary artery anomalies are rare congenital condition with an incidence ranging from 0.17% in autopsy cases to 1.2% in angiographically evaluated cases.

Methods: Retrospective review of 17,245 patients were done out of which echocardiography was used in 250 patients, Invasive coronary angiography (CAG) in 16828 patients and MDCT coronary angiography in 374 patients as primary detection modality. Indications for evaluation included angina, dyspnea and cyanosis. All patients were evaluated by transthoracic echocardiography. Invasive CAG was done in flat-panel cath-labs. MDCT CAG was done under 128 slice MDCT scan.

Results: A total of 17,245 coronary artery evaluations were done, of which 257 were found to have coronary artery anomalies at a prevalence rate of 1.49%. Most common anomaly detected in infantile period was ALCAPA (26.7%), coronary artery fistula (52%) in pediatric age group, anomalous high origin of coronary artery from same sinus, separate origin of LAD and LCX from left coronary sinus (29.7%) followed by anomalous origin of LCX from right coronary sinus (17.58%) in elderly population. 79.4% had benign anomalies while 20.6% had malignant coronary anomalies. Anomalous origin and course was detected in 202 patients (78.59%) and anomalous coronary termination in 35 (13.62%) patients.

Conclusions: Echocardiography adequately detected proximal coronary anomalies especially in pediatric patients. Invasive coronary angiography and MDCT coronary angiography were comparable for detection of most anomalies of coronary origin (except anomalous high origin near proper sinus). Invasive CAG was slightly better for distal anomalies like anomalous anastomosis.

Safety and efficacy of percutaneous device closure of large post-tricuspid shunts with severe pulmonary artery hypertension in pediatric patients

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Background: Transcatheter closure of large post-tricuspid shunts in patients with severe pulmonary arterial hypertension (PAH)