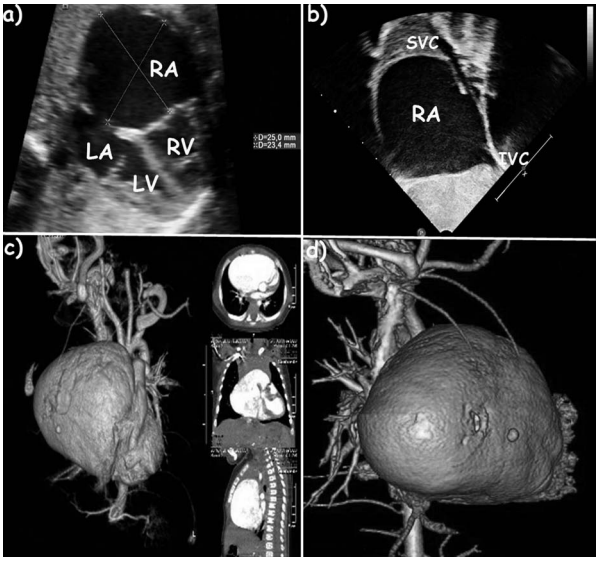
Objectives: To determine to which degree pulse oximetry screening (POS) contribute to an earlier diagnosis of isolated transposition of the great arteries (TGA) and to estimate the proportion of neonates with TGA who would benefit from a prenatal diagnosis.
Methods: Infants with TGA born in our referral area for pediatric cardiac surgery from 2003-01-01 to 2013-08-01 were identified from our surgical files and from the causes of death registry (National Board of Health and Welfare). Data on clinical presentation, diagnosis, management and outcome were collected from hospital charts.
Results: 91 cases were identified of which 34 were born in hospitals using POS. 3 were diagnosed prenatally. 57 developed early symptoms and were diagnosed before routine newborn physical examination and before POS. 7 were detected by POS, 13 at the routine newborn physical examination, 1 after that examination but before discharge and 10 were discharged undiagnosed. Five of those discharged were detected at a routine follow-up visit (one at 6 weeks of age) and 5 came to hospital because of symptoms. None of 34 born in hospitals with POS were discharged undiagnosed compared to 10 of 57 in the remaining hospitals. The age at suspicion of congenital heart disease was significantly lower in those born in hospitals with POS. 61 underwent balloon atrial septostomy (BAS), in 9 cases before 6 hours of age and in 26 cases before 12 hours. One neonate died at 4 hours of age at the referring hospital after an unsuccessful BAS. There was no further deaths at a median follow-up of 4,4y (30d-10y). 9 children had neurological symptoms at follow-up of which 3 had neurological symptoms (seizures) already before the arterial switch operation. These three neonates had severe hypoxia (20-50%) and high levels of lactate at arrival at our centre. One needed CPR immediately after arrival. BAS was performed at 6, 8 and 14 hours of age respectively.
Conclusions: Although POS prevents discharge of infants with undiagnosed TGA, at least 4 of 91 (all born in hospitals using POS) would have potentially benefited from a prenatal diagnosis to avoid death or neurological sequelae.

MP2-24
Prenatal Diagnosis And Outcome For Fetuses With Congenital Absence of The Pulmonary Valve
Babaoglu K. (1), Uzun O. (2), Deveci M. (1), Kayabey O. (1)
Kocaeli University Medical Faculty Kocaeli Turkey (1); University Hospital of Wales Cardiff Wales UK (2)

Objective: Absent pulmonary valve syndrome (APVS) is a rare congenital anomaly with its hallmark feature of a rudimentary and dysplastic pulmonary valve. In most instances, it is associated with severe dilatation of the main and branch pulmonary arteries (PA) owing to the combination of severe pulmonary stenosis and regurgitation. The aim of this study is to review the spectrum of the prenatally detected APVS and its outcome after diagnosis.
Methods: Clinical data and echocardiographic findings of 11 cases with a fetal diagnosis of APVS between 2008 and 2013 were analyzed in this retrospective two-center study. Collected parameters included: gestational age at referral; associated cardiac, genetic and non-cardiac fetal abnormalities; maximum diameters of the aortic and pulmonary annuli in addition to the main and branch pulmonary arteries.
Results: Median gestational age at diagnosis was 21 weeks. Four subtypes of APVS were observed: (1) with tetralogy of Fallot (TOF) (n = 6; 54%); (2) isolated (n = 3; 27,7%); (3) with CAVSD (n = 1; 9,9%); and (4) with VSD (n = 1; 9,9%). Ductus arteriosus was restricted in 5, absent in 3, and large in 3 fetuses. Two pregnancies were terminated. Two fetuses are still to be delivered. One fetus was stillborn who had trisomy 18. Of the six live births, one neonate died following birth due to severe hypoxia, 3 died after surgery, one remains well after operation, and the last patient is on medical follow up without operation. The presence of ductus arteriosus is not associated with survival. The genetic survey was abnormal in 27,7% of fetuses (trisomy 18 in one, and 22Q11microdeletion in two).
Conclusion: Prenatal diagnosis of the APVS is rather straightforward because of its typical features of a dilated main pulmonary and branch arteries, and color Doppler detection of severe stenosis and insufficiency of the functionally absent pulmonary valve. Outcome of antenatally APVS is poor and survival is mainly determined by the presence of respiratory symptoms due to bronchio-tracheal obstruction in this study.

MP2-25
Idiopathic dilatation of the right atrium. Report of four fetal cases.
Walter C., Carretero J.M., Bartrons J., Prada F.
Hospital Sant Joan de Déu. Barcelona. Spain.

Introduction: Idiopathic dilatation of the right atrium (IDRA) is a rare anomaly defined as isolated enlargement of the right atrium in the absence of other cardiac lesions or predisposing conditions to cause right atrial (RA) dilatation, especially tricuspid valve diseases. The clinical presentation is highly variable from asymptomatic to cardiac failure or even sudden death. It can be associated with atrial arrhythmias, thrombus formation and congestive heart failure.



Methods: We report 4 cases of IDRA of prenatal diagnosis, three children and last one is still a prenatal case. We describe the intrauterine course, the postnatal management and its short-medium term follow-up. Echocardiography of the fetus showed an abnormal 4-chamber view with an increased cardiothoracic circumference ratio at the expense of enlarged RA without other anomalies. During the routine controls the dilatation of RA persisted. None had extracardiac malformations. Postnatal echocardiography demonstrated that the RA was dilated with spontaneous echo contrast (Figure 1a-b). Although none had thrombus formation in the RA we decided initiate treatment due to a potential risk for pulmonary embolism. In all patients thrombophilia study was normal so we started thrombosis prophylaxis with salicylic acid. There has been no need for

surgical intervention so far because of the lack of arrhythmias or symptoms, although one of our cases we have found an important progression of RA diameters. This patient is 4 months old and the RA is massively dilated (diameter of 65 mmX71 mm detected with angioCT). Cardiac resonance imaging showed global hypokinesia and he is awaiting surgical decision (Fig. 1c-d).

Conclusions: Optimal management of IDRA is controversial and depends on the individual case. This disease probably is under diagnosed, reinforced by the fact we have seen four cases, but all in the last two years.

Long-term follow-up is necessary to monitor progression of RA size and occurrence of arrhythmias. Asymptomatic patients can be managed medically but symptomatic patients may require surgical reduction of the RA. Different imaging techniques including computed tomography and cardiac resonance imaging are useful to diagnose, evaluate and perhaps it may be helpful in treatment decisions. According to Paladini a Holt-Oram syndrome should be ruled out.

MP2-26

Fetal aortic valvuloplasty – how to improve outcome ?

Dangel J. (1), Debska M. (2), Kolesnik A. (3), Kraska A. (4), Debski R. (5)

Perinatal Cardiology Department, 2nd Dept. of Obstetrics and Gynecology, Medical University of Warsaw (1); Department of Endocrinology, Centre of Medical Postgraduate Education (2); Cardiac Catheterisation Laboratory, Children's Memorial Health Institute (3); Cardiology Department, Children's Memorial Health Institute (4); 2nd Department of Obstetrics and Gynecology, Centre of Medical Postgraduate Education (5); Warsaw, Poland

Introduction: Fetal balloon aortic valvuloplasty F-BAV has already been performed in few institution all over the world. The two biggest series published different outcome: 30% of bivetricular circulation (BV) in Boston and 70% in Linz. There is still not consenus what is the best treatment for neonates and infants after FBAV. Prenatal natural and after FBAV history is still far from understanding. Knowing this we started the program of fetal cardiac interventions in 2011. The objective of this study was evaluation of preliminary results of FBAV.

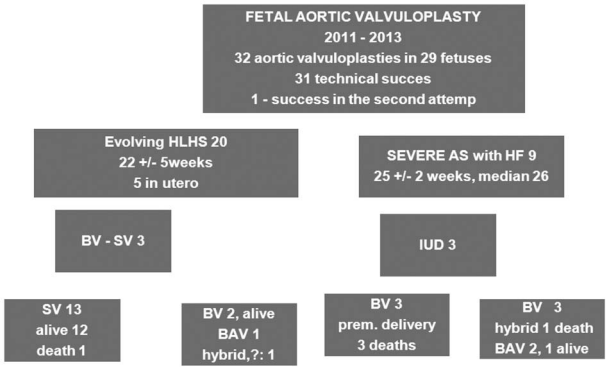
Material: Between 2011–2013 32 FBAV was performed in 29 fetuses. Fetuses were divided into two gropus: evolvingHLHS (eHLHS)-20; severe AS with heart failure-9.

Results: Aortic valve was successfully dilated in all 29 fetuses. 11 procedures were done under general anesthesia of the mother, 13 – intravenous, recent 6 – local. All fetuses had intraumbilical analgesia with fentanyl.

In 20 fetuses with eHLHS there was better flow through the aortic valve and better LV function after the procedure. In spite of this just 2 had biventrivular circulation. One was switched to BV after hybrid procedure in the neonatal period. 15 fetuses from the eHLHS group survived neonatal and early infants period. In 3 the first attempt was BV circulation, but it had to be switched to SV due to very poor LV function. One child from this group died due to severe heart and multiorgan failure.

Fetuses from the second group were in worse condition. 3 were hydropic, 3-severe LVdysfunction with closed Fo and polyhydramnion. In spite of successful procedures, 3 died in utero, 3 were born premature, 2 died without treatment,1 after 2 BAV in the neonatal period. The last after FBAV and stent placement into IAS, died in th 5th week of life after hybrid procedure. There is just one BV survivor from the second group.

Conclusions: FBAV can be successfully performed. The prenatal course after successful dilation of the aortic valve is unpredictable. Fetuses with severe heart failure are in much higher risk than eHLHS. The best postnatal treatment of this difficult patients should be the topic of international discussion.



MP3-1

Long-term fate of children operated for the hypoplastic left heart syndrome in a country with high foetal termination rate and centralized paediatric cardiovascular care

Vojtovic P., Tlaskal T., Gebauer R., Hucin B., Reich O., Selko M., Chaloupecky V., Skovranek J., Tomek V., Krupickova S., Matejka T., Hecht P., Janoušek J.

Children's Heart Centre, University Hospital Motol, Prague, Czech Republic

Objectives: To evaluate long-term results of surgical palliation for the hypoplastic left heart syndrome (HLHS) on a territory (Czech Republic, 10.5 million inhabitants) with a country-wide prenatal detection program (estimated detection rate of HLHS >90%, termination rate of detected HLHS 87%) and postnatal care centralized to one paediatric cardiovascular centre.

Methods: Retrospective analysis of all consecutive newborns with HLHS admitted between 1999-2012. Non-obligatory re-interventions were defined as those additional to the 3 stages of the Norwood pathway.

Results: From a total of 65 consecutive newborns with HLHS 13 pts. (20%) did not receive surgical treatment because of parental decision, associated anomalies or non-fulfilment of the indication criteria. 52 patients (prenatal diagnosis in 33%) were directed to the Norwood pathway (median age/weight 7 days/3.2 kg). Early/total mortality after Norwood stage I was 9.6/19% (10/52 pts.) with a significant risk factor being lower weight at surgery (Cox proportional risk per 1 gram = 0.997, CI 0.995-0.990, $p < 0.001$). Between Norwood stage I and II 15 catheter/surgical re-intervention were carried out in 13 pts. (aortic arch narrowing in 10/13). 42 pts. aged median 6.8 months underwent stage II surgery with a total mortality of 4.8% and 18 subsequent re-interventions in 13 patients. Finally, 26 patients aged median 3.9 years underwent Fontan completion with early/total mortality 0 and 8%, resp. The probability of survival at 1/5/10 years of age was 77/77/71 %. Probability of freedom from non-obligatory surgical/catheter reinterventions was 58/45/41 %. At long-term follow-up (median 7.8 years) 37/38 pts. are in NYHA functional class I or II.

Conclusions: Due to high foetal termination rate the population of live-born HLHS patients is biased towards those without a prenatal diagnosis. Despite a highly centralized care, surgical treatment of HLHS is still associated with significant mortality and morbidity. Long-term survivals, however, have an acceptable