ECOCARDIOGRAFIA FETALE E NEONATALE

P12

PRENATAL DIAGNOSIS OF VEIN OF GALEN ANEURYSMAL MALFORMATION: IMPACT OF ECHOCARDIOGRAPHIC EVALUATION ON PERINATAL MANAGEMENT

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Objectives: to describe our experience in the echocardiographic evaluation and perinatal management of prenatally diagnosed vein of Galen aneurysmal malformation (VGAM).

Background: VGAM is a congenital vascular malformation that comprises 1% of all pediatric congenital anomalies. Regardless of type of treatment, perinatal mortality and morbidity are high, often due to cardiac failure.

Methods: the cardiology department registry was retrospectively searched for all patients born between January 2008 and December 2012 with a prenatal diagnosis of VGAM. Variables assessed were gestational age (GE) at diagnosis and at time of delivery, prognostic echocardiographic indices, type of delivery, clinical condition at birth and outcome.

Results: 12 cases of VGAM were diagnosed at a median GE of 33 weeks (± 2.7). Cardiomegaly was detected in all cases. Seven fetuses showed severe cardiomegaly with dilated superior vena cava (SVC) and retrograde aortic flow. Four of them had also retrograde A wave in the ductus venosus (DV). They were all delivered with caesarean section (CS) at mean GE of 36.6 weeks (±1.3). At birth 3 of them presented with high output cardiac failure and died within few days without any invasive treatment. The other 4 underwent early endovascular treatment (between one week and 2 months of life). They all have a stable neurological status except one who had cerebral hemorrhage and hydrocephalus after embolization. Among patients with moderate cardiomegaly we found dilated SVC in 3 cases and retrograde aortic flow in 2. There were 4 vaginal deliveries and 1 scheduled SC at a mean GE of 38.6 weeks (±0.5). They were well compensated at birth and underwent one or more embolization later in life (between 2 and 5 months) except one who showed spontaneous resolution of the lesion. They are all doing well with normal neurocognitive outcome.

Conclusions: we confirmed that cardiomegaly is a warning although not specific sign of VGAM in the third trimester of pregnancy. The more severe is the cardiomegaly the worse is the postnatal outcome. Although VGAM still carries a very poor prognosis, prenatal diagnosis allows to improve perinatal management decreasing the incidence of cardiac failure and increasing the chance of successful treatment.
LONG TERM FOLLOW-UP OF FETAL CASES WITH TRICUSPID VALVE ANOMALIES

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Objectives of the study: retrospective-prospective study of the characteristics and long-term outcomes of fetal cases with tricuspid valve anomalies, aiming to assess negative prognostic factors.

Material and methods: The echocardiographic data and long term outcome (6m-27yrs) of 41 fetuses diagnosed between 1986 – June 2012 as Ebstein (Ebst) or non-Ebstein anomaly (NE), at 21-37 week’s gestation (wg), median 29, were analysed. Nineteen cases had Ebst and 22 NE, one with mitral dysplasia and 1 with coarctation. Two fetuses had extracardiac anomalies, 3 Ebst had familial history of congenital heart disease, 1 mother was taking lithium and 1 gardenal. Following variables were compared by means of Wilcoxon and Chi-square tests in cases who died and in survivors: grade of tricuspid regurgitation (TR), Celermajer index (CInd), cardiothoracic ratio (CR), fetal hydrops (FH), pulmonary stenosis/ataresia (PS, PAtr).

Results: Echocardiographic features: 13/19 fetuses with Ebst had a moderate-severe displacement of the TV and moderate-severe TR at presentation, 3 had pulmonary stenosis (PS) and 5 pulmonary atresia (Patr), 13/22 cases with NE had severe TR, 3 had PS and 5 PAtr. Seven had fetal hydrops (FH)–5 Ebst, 2 NE. Outcome: 3 cases opted for the termination of pregnancy, 3 Ebst died in utero (2 FH, 1 supraventricular tachycardia). Thirty six cases were delivered at 31-39 wg. Six neonates died spontaneously at 1-7days (3Ebst, 3NE). Five neonates with Ebst and 6 with NE were operated: 7 died, 4 NE survived. One Ebst died late at 3 yrs for resistant complex arrhythmias and severe worsening. Total mortality was 17/38 cases (44,7%), 12/17 Ebst (70,6%), 5/21 NE (23,8%).

Five cases that died had FH, 2 severe arrhythmias, all a higher grade of TR, CInd>1, CR >0.65 and 9 had PAtr; the variables TR, CInd and PAtr were highly significantly different with respect to the survivors (p=0.002-0.006). Twenty one cases with milder forms are alive at 6m-27 yrs, stable or improved (5 Ebst, 16 NE).

Conclusions: Our data confirm a relevant mortality of severe tricuspid valve anomalies diagnosed in utero, main negative prognostic factors being the grade of TR, CInd and PAtr. Milder forms of both variants stabilized after birth.
INCIDENCE OF MAJOR CONGENITAL HEART DISEASES IN TWIN GESTATIONS IN OUR EXPERIENCE OF FETAL ECHOCARDIOGRAPHY: 2006-2012 YEARS

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Background: Twin pregnancies are known to be affected by higher rates of congenital malformations than singleton gestations. Recent literature points to a possible increase in the Congenital Heart Disease (CHD) prevalence among monochorionic (MC) twin gestations.

Aim: to estimate the incidence and the risk of major CHDs in twins referred to our institution for fetal echocardiography.

Methods: A retrospective analysis was performed in 4469 pregnant who underwent fetal echocardiography between 2006 and 2012.

Results: Two-hundred fifty twins were registered, including 63 (25%) conceived with assisted reproductive technologies (ART) and 187 (75%) naturally conceived. The group with spontaneous conception included 80 Dichorionic (DC) (43%) and 107 MC (57%) pregnancies. In the ART group, 58 were DC (92%) and 5 were MC (8%). The incidence of CHD in naturally conceived twins was 3.2% (6/187) while in ART twins was 7.9% (5/63). Furthermore, the incidence of CHD increased in DC twins compared to MC (82% vs 18%; p=0.05) for both spontaneous conception (67% vs 33%) and ART (100% vs 0%).

Conclusions: Our results showed an higher recurrence of CHD in DC twins, supporting ultrasound screening in twins pregnancies regardless of chorionicity.
BICUSPID AORTIC VALVE IN PEDIATRIC AGE: RETROSPECTIVE STUDY OF 158 CONSECUTIVE CASES FOLLOWED IN A SINGLE CENTER

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Introduction: Bicuspid aortic valve (BAV) is a common congenital malformation, with an incidence of 1.5 – 2.0% in the general population. We present a retrospective study of 158 consecutive cases diagnosed in pediatric age in our Center.

Materials: From January 1982 to December 2012, in our Center, a BAV was diagnosed in 158 patients, 105 males and 53 females (M / F = 2:1). The mean age at diagnosis was 4 years and 8 months. A family history of BAV was found in 4%. The mean follow-up was 12 years.

Comorbidity: In 45/158 patients (28%), BAV was associated with other congenital heart defect: coarctation of the aorta in 25 cases, mitral regurgitation in 6, interatrial septal defect in 5, interventricular septal defect in 4, patent ductus arteriosus in 3, mitral stenosis in 2. A chromosomal abnormality or genetic syndrome was found 14 patients (9%).

Valve Morphology: valve morphology was classified on the basis of echocardiographic findings and, in patients operated, on anatomical description: R-N type BAV was found in 50% of patients, R-L type in 47.6% and L-N type in 2.4%.

Valve function: regarding valve function at the time of diagnosis, 29% of patients had a normal functioning valve, 25% had isolated valvular insufficiency, 16% isolated stenosis, and 30% steno-insufficiency.

Ascending aorta dilatation: during the follow-up 43/158 patients presented an aortic root diameter at upper limits o over compared to the expected values for body area.

Treatment: during the follow-up 28/158 patients (18%) required surgery or interventional procedure related to valve dysfunction valvulotomy in 8 cases, balloon valvuloplasty in 17 cases (16 percutaneously and 1 intraoperative) Bentall procedure, Ross intervention, ascending aorta substitution + surgical valvuloplasty (for acute dissection of aorta in Marfan syndrome) in 1 case respectively.

Reintervention: 3 patients had redo operation, performed by Ross procedure in 2 cases and Bentall intervention in 1 case.

Mortality: No mortality throughout the follow – up period.

Conclusions: Our results showed that, in a population of 158 consecutives pediatric patients, 18% required a therapeutic procedure (surgical or interventional), percentage rised a 20% considering the cases of redo operation. Valvulotomy and percutaneous valvuloplasty were effective in procrastinating more radical procedure later out of pediatric age. Often, aortic root presents already in pediatric age different grades of dilatation.

Bibliography
ECHOCARDIOGRAPHIC TDI ANOMALIES IN PEDIATRIC RHEUMATIC DISEASES: POSSIBLE EARLY MARKERS OF CARDIAC INVOLVEMENT?

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Cardiovascular involvement is well known in adult patients with rheumatic diseases, but very few studies have investigated pediatric population (PRD).

OBJECTIVE
To look for echocardiographic markers of cardiovascular involvement in a consecutive series of PRD patients.

METHODS
We analyzed 55 patients affected by PRD with complete echocardiography including Doppler tissue Imaging (TDI). We performed the TDI analysis in the medial (MM) and lateral (LM) mitral annulus and in the lateral tricuspid annulus (LT). We measured systolic (S), early diastolic (E), late diastolic (A) peak velocity, ejection time, isovolumic contraction time (IVCT), isovolumic relaxation time (IVRT) and diastolic time.

Our sample included 42 patients (17 males and 38 females) with juvenile idiopathic arthritis, 6 systemic lupus erythematosus, 2 dermatomyositis, 3 Behçet's disease, 1 sacroileitis, 1 juvenile scleroderma. Mean age at evaluation was 13.9±7.9 years, mean disease duration 6.7±6.9 years. Only 5 patients had clinical and laboratory signs of active disease.

Each patient was matched with a healthy control (CG). The CG was composed of 53 subjects (19 males and 34 females, age 13.5±7.6 years).

RESULTS
No differences in M mode and B mode echocardiographic left ventricle main parameters were observed between PRD and CG. We found a lower TAPSE in PRD than in CG (21.7±4.1 vs 24.1±3.8; p=0.004). Valvular abnormalities in PRD were higher than in CG: 20 cases of more than trivial tricuspid regurgitation; 8 mitral valve insufficiency, 4 mitral prolapses, 2 bicuspid aortic valves, while in the CG we found tricuspid regurgitation in 13 cases, aortic insufficiency in one case. Three patients had a small pericardial effusion. Mitral TDI peak velocity was similar between PRD and CG (MM: S 8.08±1.36 cm/sec vs 8.22±1.38 cm/sec; E 13.13±2.43 cm/sec vs 13.73±2.00 cm/sec; A 6.39±1.55 cm/sec vs 6.21±1.52 cm/sec - LM S 10.44±2.51 cm/sec vs 9.77±2.39 cm/sec; E 17.88±3.32 cm/sec vs 18.48±2.80 cm/sec; A 7.17±1.93 cm/sec vs 6.62±1.61 cm/sec). There was no difference between PRD and CG LT S (12.43±2.03 cm/sec vs 12.45±1.82 cm/sec) and LT E (12.78±2.21 cm/sec vs 12.59±1.79 cm/sec) while A was significantly higher in PRD than in CG (9.72±2.49 cm/sec vs 8.41±2.26 cm/sec; p=0.009). Among TDI time measurements almost all parameters showed significant difference between PRD and CG (MM IVCT 66.1±18.7 ms vs 57.5±14.6 ms, p=0.006; MM IVRT 76.1±16.8 ms vs 50.9±9.2 ms, p=0.001; LM IVCT 60.9±18.3 ms vs 53.4±14.1 ms, p=0.03; LM IVRT 65.3±13.8 ms vs 45.5±12.8 ms, p=0.001; LT IVCT 44.9±16.7 ms vs 37.6±13.1 ms, p=0.01).

We investigated the correlation between echocardiographic findings and clinical parameters such as active disease score, use and duration of drugs: a positive correlation was found only between methotrexate therapy and LT IVRT.

CONCLUSIONS
Save from mild valvular abnormalities in the early stages of PRD, we did not observe systolic and diastolic dysfunction, not even with TDI analysis. Small number and clinical heterogeneity of patients in our series are probably important limitations.
However, the discrete decrease of TAPSE and the increase of IVCT and IVRT may be markers of early structural abnormalities, such as hypertrophy or interstitial fibrosis and impaired myocyte relaxation. These patients might have increased risk of developing clinical cardiac disease in adulthood and thus might benefit from serial cardiac evaluations to highlight subclinical alterations that could require specific therapy.
MATERNAL GESTATIONAL DIABETES MELLITUS AND FETAL HEART: FOLLOW UP OF 78 CASES IN A THIRD LEVEL CENTER

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Introduction: Poorly controlled gestational diabetes mellitus (GDM) complicates 1.5% of pregnancies. Its effects on fetal heart are well known.

Aim: Aim of our study was to describe the incidence of fetal heart hypertrophy and fetal congenital heart disease in a population of fetuses referred for fetal echocardiography in our third level center (AORN dei colli - AO Monaldi, Second university of Naples) for poorly controlled GDM.

Population: Since January 1995 to April 2013 we practiced 8143 fetal heart scans, 78/8143 (1%) were referred for poorly controlled GDM. Mean gestational age at fetal echocardiography was 20.3 ± 1.5 weeks. We excluded mothers affected by type I diabetes mellitus. We diagnosed 1219/8143 (16%) consecutive fetuses with CHD. 15/1219 (1%) were in fetuses whose mothers were affected by poorly controlled GDM.

Results: GDM fetuses’ cardiac ventricular walls were thicker after the period of 28-34 weeks in 32/78 (41%) cases. Hypertrophy regressed during the first three months of life in 27/32 (84%) neonates. 5/32 (16%) children are still affected by hypertrophyc cardiomyopathy at a mean age of 7±2 years.

Moreover, we observed 15/78 (19%) CHD (median gestational age: 21 weeks). 7/15 (46%) atrial septal defects; 5/15 (33%) ventricular septal defects; 1/15 (7%) transposition of the great arteries; 1/15 (7%) aortic valve stenosis and 1/15 (7%) pulmonary atresia with VSD.

Among the 78 fetuses, we did not observe fetal deaths, so 100% were alive at birth. Deliveries occurred at a mean gestational age of 38.6 weeks. Survival was 99% (77/78): 1/78 (1%) died at 14 days of life after switch operation for transposition of the great arteries.

Conclusions: Poorly controlled gestational diabetes mellitus is a relatively rare but potentially serious condition. In our population, 47/78 (60%) of mothers delivered heart-affected babies: 15 (32%) CHD and 32 (68%) affected by ventricular hypertrophy.
PRENATALLY DIAGNOSED FETAL TACHYARRHYTHMIAS AND PERINATAL MANAGEMENT: EXPERIENCE OF OUR CENTRE


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Background: According to 2010 SIEOG Guideline, persistent fetal tachyarrhythmias (FT), defined as fetal heart rate at about 180-200 beats/minute, are one of the referral indications for fetal echocardiography. The latter has to evaluate type of arrhythmias (even if exact definition may be difficult), signs of fetal heart failure and the rare, but possible, associated congenital heart diseases (CHDs). Most of FT have a good prognosis and have spontaneous resolution, without serious complication for maternal/fetal health. On the other hand, depending on time of onset, duration of tachyarrhythmia and median heart rate, fetal heart failure and non-immune hydrops are rare severe occurrences that need admission and immediate therapy. The majority of FT are idiopathic, probably due to immaturity of myocardium and electrical tissue of the heart. Some FT are caused by maternal (fever, chorioamnionitis, use of B simpatico-mimetic agent) or fetal causes (such as viral myocarditis, CHDs, true rhythm disturbance). Echocardiographic monitoring is mandatory in case of persistent FT. Therapy may be started depending on fetal heart rate, type of arrhythmias and the presence of signs of heart failure. Optimal transplacentar treatment strategy is still unknown and has to be evaluated case by case, considering maternal/fetal consequences, pharmacological and pharmacokinetic properties and the hypothetical teratogenic effects of the therapy. Maternal/fetal wellness has to be closely monitored during pregnancy in case of antiarrhythmic drug use. Postnatal follow up is indicated because of the risk of recurrence and in order to define prognosis and appropriate management.

Aims: We reviewed perinatal management, efficacy of treatment choice and postnatal follow up of FT occurred in our Centre between 2009 and 2013.

Methods: Prenatal data consisted in number of fetal echocardiographies referred for arrhythmias, number of studied pregnancy, gestational age at the time of the diagnosis, number of echocardiographic control exams made for single pregnancy, type of arrhythmias, heart rate, and sign of fetal heart failure, therapeutic management and maternal/fetal complications. They were all collected retrospectively from the fetal echocardiography archive. Postnatal information concerning gestational age at birth, Apgar score, body weight, results of further cardiologic postnatal examinations, therapeutic strategy, years of follow up were collected retrospectively thanks to pediatric and pediatric cardiologist archive. Median value was used to analyze some of the data.

Results and Conclusions: Between 2009 and 2013, 94 fetal echocardiographies referred for FT were performed (median value 18, 8/years) on 45 pregnancies (median value 9/years). Persistent FT were diagnosed in 11 cases (median value 2, 2/years). Median gestational age at the time of prenatal diagnosis was 31 weeks of amenorrhea. For each diagnosed FT, 4 fetal echocardiographies had been performed on average during pregnancy. In two cases, fetal hydrops were present at diagnosis. Nine out of 11 patients received efficacious transplacentar therapy. Two patients (who are not still born) did not receive therapy. No one died during pregnancy. Three cases are still not born. Four cases had no more postnatal arrhythmias and did not receive postnatal therapy. Four cases needed pharmacological/electrical therapy in the perinatal period: two of them are still receiving pharmacological treatment, while the other two needed no more drugs. WPW syndrome was diagnosed in one case. PFO was present in 4 cases. In conclusion, FT are rare but potentially dangerous heart rhythm disturbances that can precipitate in fetal heart failure. Prenatal diagnosis of FT permits correct perinatal management of the severe cases and it allows correct definition of prognosis of non-severe cases.
BRONCHOPULMONARY DYSPLASIA ASSOCIATED WITH PULMONARY HYPERTENSION. THERAPEUTIC OPTIONS: OUR DATA IN PRETERM INFANTS

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BACKGROUND: Bronchopulmonary dysplasia (BPD) is a chronic lung disease due to immaturity, barotrauma and oxygen toxicity, developing in premature infants that requires treatment with oxygen and/or positive pressure ventilation. Its incidence may rise 47% in preterm infants aged <30 weeks (wks)[1]. Pulmonary hypertension (PH) can complicate the course of BPD and contributes to late morbidity and mortality during infancy [2]. Different therapeutic strategies have been adopted to prevent the consequences of a severe PH, especially nitric oxide (NO).

Sildenafil, a phosphodiesterase type 5 (PDE5) inhibitor, has been lately introduced in the treatment of BPD associated with PH. Long term treatment with sildenafil has been related to an improvement of the pulmonary hemodynamic but it seems to have no effects on gas exchanges [3].

OBJECTIVE: That being so the small number of data on the use of sildenafil in neonates, we report our experience, focusing on the demographic characteristics of our patients.

STUDY DESIGN: We considered 12 consecutive newborns who had presented severe respiratory distress syndrome (RDS) evolved in BPD (gestational age (GA) < or =30 wks, weighing<1500 grams) and who were followed in our Pediatric Cardiology Unit from 2009 to 2012.

RESULTS: All the newborns had a GA < or =30 wks (mean 26.8 ±1.9 wks) and weight<1500 g (mean 856 ±286 g). Eleven on 12 (92%) were born from Caesarean, 11 on 12 (92%) had received antenatal steroids dose. All the infants had severe respiratory distress and received invasive mechanical ventilation. The mean FiO2 value was 45% for average time of 10 days (± 8) then it was gradually lowered to 21% according to the clinical and laboratoristic conditions of the infants. Two newborns (17%) received High Frequency Oscillatory Ventilation (HFOV). The average duration of the ventilation was of 48 ±27 days, the average timing of the hospitalization was of 126 ±57 days. All the 12 infants developed a late onset PH with average right ventricular pressure of 43 ±15 mmHg and received hydrochlorothiazide, spirinolactone and furosemide according to the prevention and treatment protocol of the BPD. The 6 patients with higher right ventricular pressure values, two of whom had previously received NO therapy with only partial benefit, were treated with oral sildenafil at a starting dosage of 0.5 mg/kg to a maximum of 2 mg/kg/dose every 6-8 hours. The therapy with sildenafil was started on average at 58 ±18 days of life and was continued on average for 190 ±135 days. All these 6 infants showed significant lowering of right ventricular pressure (mean value pre-treatment: 56 ±9 mmHg; mean value post-treatment 25 ±4 mmHg; mean pressure variation= 32 ±10 mmHg; p=<0.001). We didn’t observe any adverse side effect. A patient who had not received either sildenafil or antenatal steroids died at the age of two years. Eight infants on 12 (67%) needed oxygen home care. Of these 5 patients had received sildenafil and 3 had not received it.

CONCLUSION: In our series the sildenafil therapy showed a complete efficacy and tolerability so that we consider it as a part of treatment program of lung disease with PH in preterm infants. Our short experience needs, obviously, further data to confirm the usefulness of this drug in neonatal population.

REFERENCES
ISOLATED III DEGREE ATRIOVENTRICULAR BLOCK IN FETUSES AND NEONATES: A RETROSPECTIVE STUDY OF 12 PATIENTS

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Introduction: Isolated complete atrioventricular block (icAVB) is a rare but potentially lethal condition in which the effects of maternal steroid treatment on outcome is unclear.

Aim: The objective of this work was to study risk factors associated with death and the influence of steroid treatment on outcome in a population of fetuses referred for fetal echocardiography for icAVB in our third level center (AORN dei colli - AO Monaldi, Second university of Naples).

Population: Since January 1995 to April 2013 we practiced 8143 fetal heart scans. Among these, we diagnosed 1219 (16%) consecutive fetuses with CHD. We retrospectively collected the data concerning 10/1219 (0.08%) fetuses diagnosed with third-degree atrioventricular block. 7/10 (70%) patients were referred for a suspicion of icAVB on obstetric scanning. 3/10 (30%) were referred because of maternal antibodies status. Mean gestational age at diagnosis was 19.3 +/- 1.4 weeks. In 100% of pregnancies we documented antibody status: the icAVB was associated with maternal anti-Ro/SSA and anti-Ro/SSB antibodies. Moreover, we observed 2 neonates with postnatal diagnosis of icAVB.

Results: 12 cases of icAVB: 2/12 (17%) with postnatal diagnosis and 10/2 (83%) diagnosed in utero.

Outcome of the fetal group: 7/10 (70%) were treated with high dose corticosteroids (Prednisone: 1.5 mg/Kg/die) for a median of 10 weeks. 3/10 (27%) were not treated because of the absence of maternal symptoms and the good fetal heart rate (100 bpm). These cases were the three referred only for maternal antibodies status.

We observed 2/10 (20%) fetal deaths [mean gestational age: 32 weeks] due to severe heart failure. These fetuses had presented fetal hydrops and a mean heart rate 50 bpm, in spite of Prednisone.

8/10 (80%) were alive at birth: 3/8 (38%) had not been treated and 5/8 (62%) had received maternal Prednisone.

Deliveries occurred at a mean gestational age of 35.3 weeks. Survival in the neonatal period was 88% (7/8): 1/8 (12%) died at 3 days of life for severe heart failure. Variables associated with death were gestational age at diagnosis <20 weeks, ventricular rate <50 bpm and fetal hydrops.

1/8 (12%) had an epicardial pacemaker by 1 year of age (at 8 months), she is now alive and well. 2/8 (24%) underwent pace-maker implantation after the year of age: 1 when he was aged 2 (epicardial device) and 1 when he was aged 8 (endocardial device), they are alive and well. In 1/8 (12%) the icAVB spontaneously converted in II degree AVB. 3/8 (38%) are alive and well in natural history. The mean follow up is 7±3 years.

Outcome of the neonatal group: 2/2 (100%) are alive and well in natural history.

Conclusions: Isolated complete atrioventricular block in the fetus is a rare but potentially lethal condition in which the effect of steroid treatment on outcome is unclear.

Fetal risk factors associated with a poor outcome were gestation <20 weeks, ventricular rate <50 bpm, hydrops, and impaired left ventricular function.

On the other hand, icAVB when diagnosed in postnatal life tend to have a better outcome.
A SURPRISE DURING A ROUTINE ECHOCARDIOGRAPHY OF A BICUSPID AORTIC VALVE

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Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly (1 per 300,000 live births). In this condition the left coronary artery usually arises from the main pulmonary artery and the time of onset of symptoms depends on the collateral circulation from the right coronary artery (RCA). Typically the clinical signs of myocardial ischemia and heart failure occur in the first few months of life when the pulmonary resistances fall.

We report the case of a five year old girl who came to our clinic for a periodical cardiologic check for a bicuspid aortic valve diagnosed shortly after birth. She presented a normal physical and mental development and she was completely asymptomatic. He also practiced a sport activity (dance) without manifesting reduced functional capacity or shortness of breath.

Physical examination showed a little systolic murmur and no signs of heart failure. The electrocardiogram was normal and no signs of myocardial ischemia were present.

Echocardiography showed normal size and function of the left ventricle and the absence of mitral regurgitation. The bicuspid aortic valve did not present stenosis or regurgitation. The right coronary artery, regularly originating from the right coronary sinus, had a lumen dilated (5 mm at the ostium). The left coronary artery was not detected in the left coronary sinus.

The color Doppler detected prominent coronary collaterals mapping an abnormal diastolic flow signal in the myocardium of the interventricular septum. The origin of left coronary artery was not found in the main pulmonary artery, but a diastolic flow signal was present in the right pulmonary artery (RPA). The coronary angiography confirmed the anomalous origin of the left coronary artery from the RPA and showed also its filling by extensive collateral circulation from RCA, with coronary steal into the RPA. Surgical correction was indicated for the risk of sudden cardiac death and was performed soon after diagnosis. The left coronary artery was reimplanted into the aorta and the postoperative course was good. Now, eight months after surgery, the child is well and she started practicing a little sport.

Presentation of ALCAPA in older asymptomatic children is unusual but we must emphasize that sudden cardiac death can occur as the first manifestation, particularly during exercise. In fact, the limited coronary reserve during an increased myocardial oxygen demand for stress could cause fatal ventricular arrhythmias. Surgical correction should not be delayed after diagnosis. The creation of a two-coronary-artery system, in particular direct reimplantation of the left coronary into the aorta, is currently the operation of choice, with good long-term prognosis.

This is a very rare case of origin of the left coronary from the right pulmonary artery in association with bicuspid aortic valve.

This report shows how much attention is required in a pediatric echocardiography exam; we believe that the origin of the coronary arteries should be systematically searched.
A RARE CASE OF CONGENITAL LEFT VENTRICULAR ANEURYSM

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Congenital heart aneurysms and diverticula are rare malformations that can occur in isolation or in combination with other congenital heart diseases (CHDs), vascular and thoraco-abdominal deformities. As few cases are described, little is known about epidemiology, etiology and natural history. Treatment strategy has to be individually defined.

Until now, the only case that occurred in our Centre was diagnosed at 22 weeks of gestational age (ga) during a fetal echocardiography performed because of suspicion of CHD at second trimester obstetric screening. Large normokinetic apical aneurysm/diverticulum of the left ventricle with preserved basal systolic function was described. Fetal life threatening arrhythmia (very frequent supraventricular ectopic beats, sometimes organized in runs of supraventricular tachyarrhythmia with median heart rate at about 200/min), without signs of heart failure, set in at 34 of ga. Transplacental therapy with digossin and flecainide was started. Because of maternal electrocardiographic alterations, it was then discontinued in two days. No more fetal arrhythmias occurred. The maternal-fetal wellness, the dimension of the aneurysm, the systolic function of the heart, the presence of pericardial effusion and fetal cardiac rhythm were strictly monitored during gestation through eight fetal echocardiographies. Obstetric medical history was collected, (i.e. theratogenic agent exposure, other extra-cardiac malformations, previous familiar CHDs). Multidisciplinary prenatal counseling was organized before delivery to explain diagnosis and prognosis to the parents. Normal labor happened at 39+3 weeks of ga without complication. Postnatal echocardiography confirmed prenatal diagnosis. No complications occurred during the first hospital stay. The baby was then re-admitted to hospital because of atrial flutter, moderate reduction of systolic heart function and mild mitral regurgitation and he is still undergoing therapy. Follow up is still ongoing. Other more invasive diagnosis examinations may be necessary in order to better define diagnosis and prognosis of that CHD.

Multidisciplinary approach is mandatory to allow correct and complete maternal-fetal and perinatal care.