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Prenatal diagnosis of major aortopulmonary collateral arteries

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Abstract

Background: The presence of foetal major aortopulmonary collateral arteries (MAPCAs) is associated with adverse outcome, therefore early diagnosis is essential.

Aim: To evaluate the usefulness of foetal echocardiography in the diagnosis and evaluation of MAPCAs in foetuses with pulmonary atresia, as well as to assess the effects of prenatal diagnosis on the management of neonates with pulmonary atresia.

Methods: From 11,678 examined foetuses, we retrieved 15 cases of patients with MAPCAs and congenital heart defects which had been diagnosed by foetal echocardiography (1994–2008), using 2D echocardiography + color-Doppler (CD) + pulsed Doppler (2DD) and spatio-temporal image correlation (STIC) techniques. In 13 patients, MAPCAs were confirmed after birth based on angiography.

Results: In all cases, vessels corresponding to MAPCAs were visible in longitudinal view with CD, and in three cases were additionally confirmed by STIC technique. In nine cases one, in four cases two, and in two cases three MAPCAs were suspected. In two cases, MAPCAs were not confirmed after birth; one due to misdiagnosis secondary to aberrant right subclavian artery, and one because of abnormal ductus arteriosus course coexistent with right aortic arch.

Conclusions: In foetuses with pulmonary atresia, it is possible to find MAPCAs with current technology (both 2D + CD, power angiography and real time-3D echocardiograophy [4D]). The differential diagnosis (MAPCAs or other vessels) should be included. Although prenatal diagnosis does not change the obstetrical management, it is important information for a paediatric cardiologist. Early neonatal angiography might be of great value not only in confirming MAPCAs, but also in performing cardiac intervention and in some cases preventing future heart failure.

Key words: foetal echocardiography, MAPCAs, pulmonary atresia

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INTRODUCTION

The prognosis and management of a group of patients with pulmonary atresia (PA) mainly depends on the nature of the pulmonary vascular supply. Three main categories of pulmonary vascularisation have been described: Type I — all bronchopulmonary segments are supplied by the ductus arteriosus to the main pulmonary arteries with confluent left and right pulmonary arteries; Type II — some lung segments are supplied by collaterals, others by the pulmonary arteries; Type III — all segments are supplied only by collateral vessels with non-confluent, small or non-existent pulmonary arteries.

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The presence of diverse pulmonary vascularisation of the lung by aortopulmonary collaterals has serious clinical implications [1]. Different sources of pulmonary blood flow can result in abnormal vascular development, with the risk of pulmonary hypertension developing in different pulmonary segments [2, 3]. In those with poorly developed pulmonary arteries and restrictive pulmonary blood flow, cyanosis is the main clinical manifestation, while in those with a large number of wide aortopulmonary collaterals, early heart failure is manifest [4]. Current ultrasonographic diagnostics should allow for the identification of aortopulmonary collaterals before birth [5]. The aim of our study was to report on the utility of foetal echocardiography in the diagnosis and evaluation of major aortopulmonary collateral arteries (MAPCAs) in the foetus with PA. Prenatal diagnosis will allow for determination of prognosis and may speed correct therapeutic interventions directly after birth.

METHODS

Study group

From 11,678 foetuses evaluated between 1994 and 2008, we identified 15 cases with pulmonary outflow obstruction and suspicion of MAPCAs diagnosed by foetal echocardiography: 13 of them presented with PA and two with critical pulmonary stenosis. Nine foetuses had tetralogy of Fallot and PA; three had complex single ventricle; two had tricuspid atresia with PA; and one had atrio-ventricular septal defect with PA. All had post-natal evaluation in our institution. In 11 patients, MAPCAs were confirmed by post-natal cardiac catheterisation (Fig. 1). In cases where there were wide collaterals possibly leading to heart failure and pulmonary hypertension, we interventionally closed collaterals where dual supply to lung segments was identified (blood inflow into pulmonary segments from a minimum of two different sources). Collateral embolisation was performed using a Jackson-coil PDA. In one patient with critical pulmonary valve stenosis, pulmonary artery hypoplasia and the presence of numerous aortopulmonary collaterals, we simultaneously conducted percutaneous balloon pulmonary valvuloplasty (balloon catheter TYSHAK II (6 × 20 mm) and collateral embolisations.

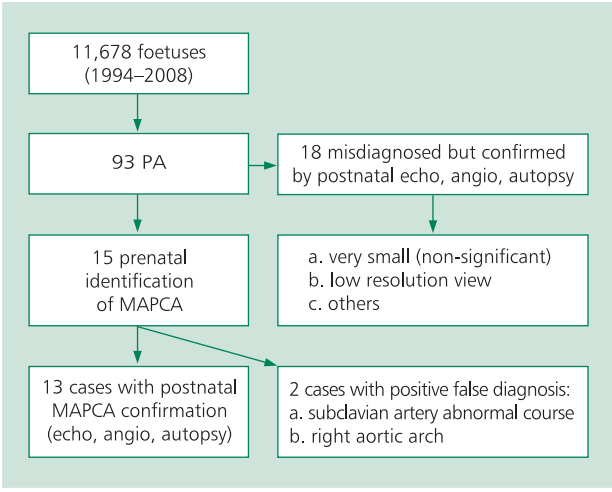

Figure 1. Foetuses retrospectively analysed; PA — pulmonary atresia; MAPCA — major aortopulmonary collateral artery; echo — echocardiography; angio — angiography

Foetal echocardiography

The ultrasound systems used were an ATL HDI-5000 and GE 730 Voluson Expert with transabdominal probes convex type C5 — 2 MHz and C7 — 4 MHz, cardiologic probe P4 — 2 MHz. A consistent standard technical approach to cardiac anatomy and function was applied in all cases: short- and long-axis scans of the intracardiac anatomy, aorta and pulmonary arteries were obtained. Colour Doppler (CD), pulsed-wave, and continuous wave were used as well. Echocardiographic recordings were retrospectively analysed from videotapes or digital media. Gestational age at the time of initial diagnosis and gestational age at the time of MAPCAs visualisation were recorded.

Angiography

After birth, all neonates had left and right-side angiography with detailed delineation of the pulmonary vasculature. Angiographic views were recorded using a uniplanar angiograph (Philips Integris CV). Aortopulmonary collateral estimation required selective angiography with use of a Judkins catheter R 3.0, vertebral and multipurpose. Pressure measurements in the pulmonary arteries and collaterals were obtained.

RESULTS

Mean maternal age was 29 ± 4.2 years. Congenital heart defects in our group of foetuses were diagnosed at mean 30.3 weeks of gestation (21–37 weeks). However, a suspicion of foetal MAPCAs was visible on foetal echocardiography later on: at mean 32.6 weeks of gestation (27–38 weeks). There were seven high-risk pregnancies (46.7%): three after in-vitro fertilisation, two based on family history (in one case the first child with common arterial trunk the other with a chest abnormality), one based on obstetric history (miscarriages at first and second pregnancy), and one with diabetes mellitus. Mean neonatal weight at the time of delivery was 2,590 ± 470 g. There was one case of intrauterine growth retardation; the others foetuses were appropriate for gestational age. There was normal mean heart size (heart area/chest area ratio) 0.39 ± ± 0.07, but in one case there was cardiomegaly (HA/CA 0.55 — case 12) [6]. There was dextrocardia in two cases and situs inversus in one case. A right aortic arch was suspected in two cases. In nine cases, there was identification of an abnormal four-chamber view with overriding aorta, ventricular septal defect and a rudimentary right ventricular outflow tract; and in six cases there was an abnormal four-chamber view due to the

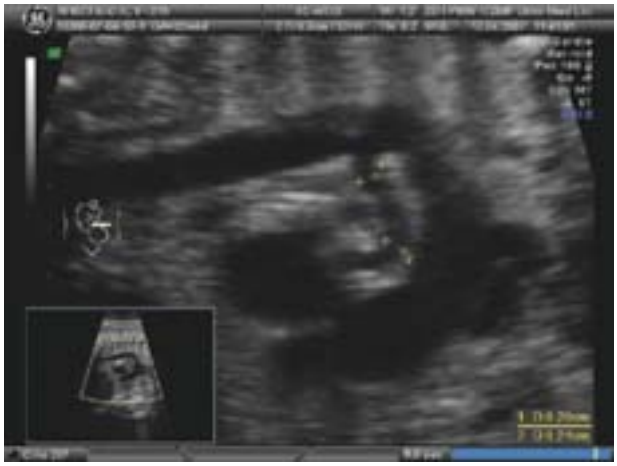

Figure 2. Foetal echocardiography. Long-axis view of MAPCA in 2D presentation. Foetus at 32 weeks and two days. Aortic arch view with MAPCA, which derived from transverse part of the aortic arch (with measurements of MAPCA)

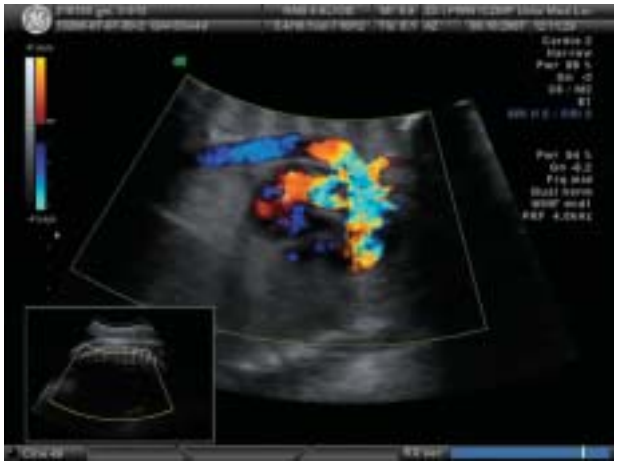

Figure 3. Foetal echocardiography (the same patient). Long-axis view of MAPCAs in color Doppler presentation

presence of a complex single ventricle (thrice), tricuspid atresia (twice), or atrio-ventricular septal defect (once). There was one false positive diagnosis of d-trasposition of great artesies in the patient number 4.

In all cases, MAPCAs were visible in the longitudinal view (Fig. 2) with CD imaging (Fig. 3). In three cases, it was also confirmed by spatio-temporal image correlation (STIC) and there was no false positive diagnosis. In nine cases, one collateral vessel was seen, in four cases two abnormal vessels, and in two cases three abnormal vessels. The vessels measured 2–4 mm in diameter on 2D echocardiography and CD. Foetal Doppler spectrum was obtained in ten cases and was similar for pulmonary flow, which can be obtained from pulmonary peripheral branches. In two cases, MAPCAs were not confirmed after birth (two cases of false positive prenatal detection). During catheterisation in these cases, an abnormal course of the subclavian artery was seen in one case (aberrant right subclavian artery) and in the other case there was an atypical course for the ductus arteriosus due to the right aortic arch.

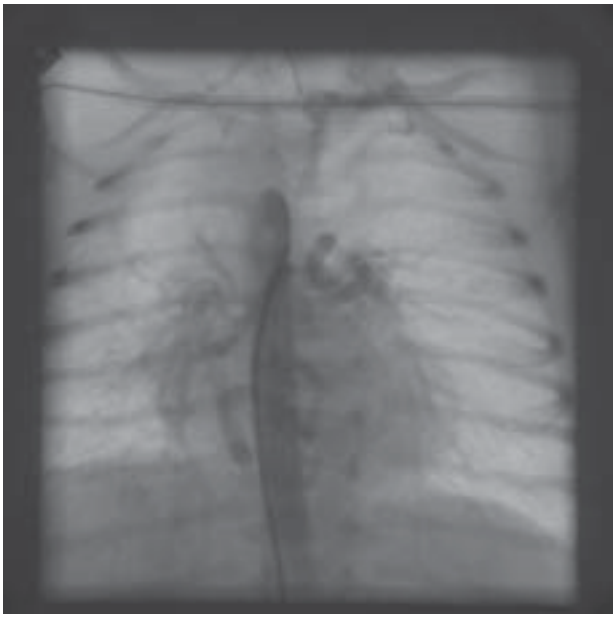

Figure 4. Angiographic view of the neonate with pulmonary atresia and ventricular septal defect. The most differential type, all segments are supplied only by collaterals (non-confluent pulmonary arteries)

In our study group, we noticed specific findings: more than one collateral visualised in foetal echocardiography can be associated with a poor postnatal prognosis (anatomical type II and III of PA). In our referral centre (Foetal Cardiology type C in Poland) we also had several foetuses without prenatal diagnosis of PA and post-natal angiographic diagnoses of MAPCAs. However, these cases were not included in this type of analysis.

In four foetuses, in addition to the main cardiac diagnosis, there was a prenatal suspicion of thymic hypoplasia. Of these, in three neonates CATCH 22 syndrome was confirmed and in one case thyroid hypoplasia was diagnosed postnatally. Cardiac catheterisation confirmed additional sources of pulmonary vascular supply in 13 patients (Fig. 4). Because of the risk of pulmonary hypertension and development of cardiac failure, two neonates had coil embolisation performed; in one case simultaneously with percutaneous pulmonary artery valvuloplasty. All the patients with MAPCAs confirmed post-natally were qualified for cardiological surgical treatment as listed in Table 1.

Table 1. Prenatal and post-natal diagnoses in a series of 15 foetuses and neonates with MAPCA

N	Gender	Gest age	Foetal dgn	No. of foetal MAPCA	Neonatal dgn	Angio type	Treatment
1	Female	37	ToF+PA	2	ToF+PA+MAPCA	II	No, MAPCA occlusion with coil, unifocalisation at four months of age
2	Male	23	SV+PA, situs inversus	1	SV+PA+MAPCA, situs inversus	III	B-T shunt to the main collateral because of low oxygen saturation due to its proximal stenosis, 46th day of life, death after surgery
3	Female	27	ToF+PA	1	ToF+PA+MAPCA	I	No surgery in four weeks of post-natal life
4	Female	24	SV/DILV+ d-TGA+PA, dextrocardia	1	SV+PA+MAPCA, dextrocardia	I	B-T shunt 10th day of post-natal life
5	Female	29	ToF+PA+Ao arch dxt	3	PA+VSD+MAPCA	III	No surgery in four weeks of post-natal life
6	Male	33	ToF+PA	1	ToF+PA+MAPCA	I	B-T shunt 21st day of post-natal life (CATCH 22)
7	Female	23	ToF+PA	1	ToF+PA+MAPCA	II	B-T shunt 28th day of post-natal life (CATCH 22)
8	Female	36	DORV/SV+VSD+PA	2	DORV+PA+ MAPCA	II	B-T shunt 31st day of post-natal life (CATCH 22)
9	Female	33	ToF+PS	2	ToF+critical PS+MAPCA	II	BVP + large collaterals occlusion using detachable coils
10	Male	31	TVA+PA	3	TVA +PA+MAPCA	II	B-T shunt 35th day of post-natal life
11	Female	36	ToF+PA	2	ToF+PA	III	No surgery in four weeks of post-natal life
12	Male	29	TVA+PA+AS+FO, RC, dextrocardia	1	SV+PA+AS, dextrocardia	II	Post-natal death, no autopsy
13	Female	31	ToF+PA	1	ToF+PA+arteria subclavia lusoris	II	B-T shunt 20th day of post-natal life
14	Male	31	AVC+PS+Ao arch dxt, DA dxt, Dandy Walker S.	1	AVC+d-TGA+PS	None	Post-natal death, autopsy (+)
15	Female	32	ToF+PA	1	DORV+PA+arteria subclavia lusoria	None	B-T shunt 36th day of post-natal life

MAPCA — major aortopulmonary collateral arteries; ToF — tetralogy of Fallot; PA — pulmonary atresia; SV — single ventricle; B-T — Blalock-Taussig; DILV — double inlet left ventricle; d-TGA — d-transposition of great arteries; Ao arch dxt — right aortic arch; VSD — ventricular septal defect; CATCH — cardiac defect, abnormal face, thymic hypoplasia, cleft palate, hypocalcemia; DORV — double outlet right ventricle; PS — pulmonary stenosis; BVP — balloon valvuloplasty; TVA — tricuspid valve atresia; AS — aortic stenosis; FO — foramen ovale; RC — restrictive cardiomegaly; AVC — atrioventricular canal; DA — ductus arteriosus

DISCUSSION

There is a great variability in the anatomy of the pulmonary arteries and the sources of pulmonary blood flow in patients with PA and ventricular septal defect [2, 4, 7]. A fully developed, normally sized pulmonary artery with confluent left and right pulmonary branches is in fact uncommon. A situation more commonly encountered is one in which pulmonary blood flow is supplied by aortopulmonary collateral arteries which usually originate from the descending aorta, the aortic arch, or subclavian artery. The number of collateral vessels can vary, as does the nature of the connection of these vessels into the lung vasculature. Collateral vessel stenosis is possible, or such vessels may be completely unobstructed [8, 9]. When there is no obstruction, these can lead to pulmonary vascular disease or the development of cardiac failure [4, 8, 9].

Prenatal diagnosis of MAPCAs is extremely important for urgent post-natal cardiac intervention and/or surgical treatment [5, 10, 11]. In most cases in our study group, we diagnosed MAPCAs using 2D echocardiography with CD methods, although in three cases the anomaly was confirmed by 4D. Some authors have suggested that 4D echocardiography with B-flow imaging and STIC, unlike 2D, can provide thorough visualisation of very small vessels and of the arterial blood supply to the lung [11]. In our group of foetuses, there were two cases of MAPCAs absent after birth. During foetal echocardiography similar to MAPCAs visualisation was registered in CD in case of abnormal course of right subclavian artery or atypical view of ductus arteriosus in course of right aortic arch [12]. It should be underlined that an atypical course of some normal structure (ductus arteriosus in right aortic arch) or an abnormal course of aortic branches may be mistaken for MAPCAs. Therefore, CD + pulsed Doppler technique may be helpful in distinguishing MAPCAs from other vessels. Nowadays in some patients there is a possibility of interventional closure (coil implantation) of MAPCAs as a protection of increased pulmonary flow and pulmonary hypertension development [4]. In cases with pulmonary vasculature hypoplasia with poor collaterals development, trials of interventional palliation with stent implantation into narrowed collaterals have been described [3, 13–15]. Due to the complexities of malformation, most patients require multistage operative treatment. The most important predictors of successful treatment in this group of patients are a sufficient degree of proper pulmonary vasculature development and a lack of pulmonary hypertension [4, 9, 16].

Conotruncal heart defects (tetralogy of Fallot, PA, common arterial trunk) are often a consequence of genetic disorders [15]. We suspected CATCH 22 syndrome in four foetuses, and in three of these patients CATCH 22 syndrome was indeed confirmed by Fluorescent In Situ Hybridisation test after birth.

Despite the progress of prenatal diagnosis, a prostaglandin infusion is necessary until the confirmation of MAPCAs at neonatal cardiac angiography.

CONCLUSIONS

In foetuses with PA, it is possible to find MAPCAs using current technology (both 2D + color Doppler, power angiography and 4D). A differential diagnosis (MAPCAs or other vessels) should be included. Although prenatal diagnosis does not change the obstetric management, it is valuable information for a paediatric cardiologist. Early neonatal angiography may be of great value, not only in confirming MAPCAs, but also in allowing cardiac intervention and in some cases preventing future heart failure.

Conflict of interest: *none declared*

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Diagnostyka prenatalna kolaterali aortalno-płucnych

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Streszczenie

Wstęp: Obecność kolaterali aortalno-płucnych u pacjentów z atrezją zastawki płucnej wiąże się z poważnymi skutkami klinicznymi. W grupie osób z dużą liczbą kolaterali szybko dochodzi do rozwoju niewydolności serca. U pacjentów ze zmniejszonym przepływem płucnym i słabo rozwiniętymi naczyniami płucnymi dominującym objawem jest sinica, podczas gdy odaortalne unaczynienie płuc może prowadzić do rozwoju nadciśnienia płucnego w poszczególnych segmentach płuc. Nowoczesna diagnostyka echokardiograficzna pozwala na rozpoznanie odaortalnego unaczynienia płuc już w okresie prenatalnym.

Cel: Celem pracy była ocena przydatności echokardiografii płodowej w rozpoznaniu i ocenie kolaterali aortalno-płucnych (MAPCAs) u płodów z atrezją zastawki płucnej oraz wpływ diagnostyki płodowej na postępowanie z noworodkiem z atrezją zastawki tętnicy płucnej.

Metody: Analizie retrospektywnej poddano 11 678 prenatalnych badań echokardiograficznych [echokardiografia dwuwymiarowa (2D) + technika Dopplera znakowanego kolorem (CD) + Doppler pulsacyjny (2DD) oraz czasoprzestrzenna korelacja obrazu (STIC)] wykonanych w latach 1994–2008 w Zakładzie Diagnostyki i Profilaktyki Wad Wrodzonych ICZMP. Wśród płodów z atrezją zastawki płucnej w 15 przypadkach uwidoczniono kolaterale aortalno-płucne w badaniu prenatalnym. U 13 z nich obecność odaortalnego unaczynienia płuc potwierdzono badaniem angiograficznym.

Wyniki: We wszystkich przypadkach naczynia odpowiadające kolateralom aortalno-płucnym uwidoczniono w osi długiej z przepływem potwierdzonym w CD, w 3 przypadkach potwierdzono je także za pomocą techniki STIC. W 9 przypadkach podejrzewano 1 kolateralę, w 4 przypadkach — 2, w 2 przypadkach — 3. W 2 przypadkach obecności kolaterali nie potwierdzono po urodzeniu. W przypadku pierwszym fałszywie dodanie rozpoznanie prenatalne było wynikiem obecności tętnicy podobojczykowej błądzącej, w drugim przypadku — nietypowego przebiegu przewodu tętniczego współistniejącego z prawostronnym łukiem aorty.

Wnioski: U płodów z atrezją zastawki płucnej jest możliwa prenatalna diagnostyka kolaterali aortalno-płucnych przy użyciu nowoczesnych technik echokardiograficznych [2D + CD + 2DD, Doppler mocy (*power-angio*), echokardiografia 3-wymiarowa w czasie rzeczywistym (4D)]. Prenatalne rozpoznanie dodatkowego odaortalnego unaczynienia płuc u płodu z atrezją zastawki płucnej nie wpływa na postępowanie położnicze, stanowi natomiast cenną informację dla kardiologa dziecięcego. Wczesna diagnostyka angiograficzna w tej grupie noworodków ma na celu nie tylko potwierdzenie prenatalnego rozpoznania kolaterali aortalno-płucnych, ale pozwala także w niektórych przypadkach na przeprowadzenie leczenia interwencyjnego w celu uniknięcia rozwoju niewydolności serca w przyszłości.

Słowa kluczowe: echokardiografia prenatalna, kolaterale aortalno-płucne, atrezja płucna

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