Abnormal or delayed development of the Area Membranacea Posterior of the brain: the Blake's pouch cyst. Anatomy, ultrasound diagnosis, natural history and outcome in the fetus.

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ABSTRACT

Objectives. Objectives of this study are: 1) To review the normal development and the pathogenesis of the posterior membranous area (PMA) in the fetal brain; 2) to define the sonographic criteria to diagnose a Blake's pouch cyst (BPC) in the fetus; 3) to review ultrasound features, associations and outcome of 19 cases of BPC seen at our center over the last 5 years.

Methods. Nineteen cases of posterior fossa anomalies with a final diagnosis of BPC were studied. The following variables were assessed: referral indication, gestational age at diagnosis, ultrasound and MRI findings, associated anomalies, natural history, pregnancy and neonatal outcome. In all cases, a transvaginal examination was performed with three-dimensional ultrasound equipment. MRI was performed in 15 cases. To confirm the diagnosis, MRI transfontanellar ultrasound or autopsy were available in all cases.

Results. Referral indications were: suspect of vermian abnormality in 11 cases (58%), other non-CNS anomaly in 8 cases (42%). Sonographically, all cases showed the following three signs: 1) normal anatomy and size of the vermis; 2) mild/moderate counterclockwise rotation of the vermis; 3) normal size of the cisterna magna. At three-dimensional ultrasound, the upper wall of the cyst was clearly visible in 11/19 cases, with choroid plexuses on the superolateral margin of the cyst roof. On follow up, the Blake's pouch cyst disappeared in 6/9 cases not undergoing termination of pregnancy at 24-26 gestational weeks remaining unaltered until birth in the remaining 3. Associated anomalies were present in 8 cases (42%) and in 5 of these consisted of or included congenital heart disease. Karyotype was abnormal in 2/12 cases (2 cases of trisomy 21) in which it was available. As for pregnancy outcome, there were 8 terminations of pregnancy (42%), 2 neonatal deaths (10%), and 9 livebirths (48%). One neonate has obstructive hydrocephaly confirmed (the BPC has disappeared),

another neonate was diagnosed with Down syndrome after birth. Neurodevelopmental outcome is normal in 7/8 cases (with the exception of the Down syndrome baby).

Conclusions. Based on the analysis of the ultrasound features, we propose that for BPC to be diagnosed in a fetus the following three criteria should be fulfilled: 1) normal anatomy and size of the vermis; 2) mild/moderate counterclockwise rotation of the vermis; 3) normal size of the cisterna magna. Furthermore, we have found that BPC can undergo delayed fenestration at 24-26 weeks in more than 50% of the cases. Finally, we have demonstrated that BPC shows a significant risk of association with extra-cardiac anomalies (heart defects in particular) and, to a lesser extent, to trisomy 21.

INTRODUCTION

The Blake's Pouch Cyst (BPC) has been considered a pathological entity deriving from an abnormal development of the Posterior Membranous Area (PMA) of the fetal brain (1-3). However, despite its first description dates back to 1900 (1,2), there is scant evidence of this pathological entity in the recent literature, with only less than 10 articles published on the postnatal appearance of BPC in infants and adults (3-8). And, despite the fact that posterior fossa cystic malformations are one of the most frequently discussed topics in fetal neurology, we could not find, to the best of our knowledge, a single series dealing with this pathological entity in the fetus from the diagnostic and clinical standpoint, the only exceptions being a pathology study (9) and another couple of interesting articles (10, 11). This is why we decided to review the basic papers dealing with the normal and abnormal development of the PMA, in order to clarify as thoroughly as possible, the origin of the BPC. Having done that, to identify sonographically adoptable criteria for such a diagnosis and, finally review our experience with the diagnosis of BPC in the fetus.

In fact, the objectives of this study are: 1) To review the normal development and the pathogenesis of the posterior membranous area (PMA) in the fetal brain, as evident from the literature published to date; 2) to define the sonographic criteria to diagnose a Blake's pouch cyst (BPC) in the fetus; 3) to review ultrasound features, associations and outcome of 19 cases of BPC seen at our center over the last 5 years.

METHODS

As far as the first objective is concerned, all the articles describing the normal and abnormal development of the PMA, cited or not cited in the limited clinical literature on the BPC were

consulted a MEDLINE search with the terms “Blake’s pouch” +/- “4th ventricle” +/- “roof” was made, and all pertinent articles reviewed (1-3, 12-16). The description of the steps leading to the normal and abnormal development of the posterior fossa and the 4th ventricle was created collating the evidences reported in this series of articles.

As for the clinical and ultrasound retrospective study, all cases of posterior fossa anomalies with a final diagnosis of BPC were retrieved from our database. The following variables were assessed: referral indication for expert targeted ultrasound (17), gestational age at diagnosis, ultrasound findings, MRI findings, associated anomalies, karyotype, natural history, pregnancy and neonatal outcome. In all cases, a transvaginal examination was performed with three-dimensional ultrasound equipment (E8, General Electrics, Milwalkee, IL). Conventional 5-9 MHz or high-frequency 6-12 MHz endovaginal volumetric transducers were used for all examinations. The ultrasound criteria adopted to diagnose a BPC **were** those illustrated above: 1) normal anatomy (including normal appearing fastigium) and size of the vermis (median section of fetal brain); 2) mild to moderate counterclockwise rotation of the vermis (median section of fetal brain); 3) normal size of the cisterna magna (median and axial sections of fetal brain); 4) inconstantly, evidence of the BPC roof within the cisterna magna (median section of fetal brain). The nomograms published by Vinals et al (18) were used to assess vermian size. Fifteen cases underwent also prenatal MRI aat diagnosis or on follow up. To confirm the BPC, a follow up transfontanellar ultrasound or an MRI were performed in cases in which the pregnancy reached term of gestation. In case of termination of pregnancy, only the normal aspect of the vermis on the median section could be assessed, due to the fact that the fluid collection often disappears at necropsy as soon as the posterior fossa is dissected.

RESULTS

1. **Development of the Posterior Membranous Area of the fetal brain.**

    a. **Normal posterior membranous area (PMA) development: the Blake's pouch** (1,2, 12-16). The *Blake's pouch* represents an evagination of the posterior membranous area (PMA), one of the two components of the rhomboencephalic roof (the other being the anterior membranous area – AMA) which develops at the Carnegie stage 14 of the embryonic development. These two components – AMA and PMA - are separated by a transverse vascular fold, the *plica choroidea,* which invaginates into the 4th ventricle and represents the first evidence of the choroid plexus, which will eventually appear in the 4th ventricular roof by stage 19 (48-51 days post-ovulation, gestational week 7, 18-20 mm). The cerebellar vermis will develop from the rhombic lips of the AMA due to intense cell proliferation, and this will lead the forming choroid plexus to be displaced caudally, cranially to the PMA. Until this time, it is assumed that there are no communications between the 4th ventricle and the cisterna magna; in fact, it seems that in most cases the lateral apertures of Luschka appear much later: between the 14th-17th week of gestation according to some authors (12-15), or even not before the 26th week according to others (16); in some 20% of cases they do not open at all (12-15). The median Magendie foramen seems to open from the 9th to the 10th gestational week (see below). If that is the case, then the only way to explain the presence of cerebro-spinal fluid in the sub-dural spaces is through cavitation. At 7 post-ovulation weeks (9 gestational weeks), an evagination begins to appear on the roof of the 4th ventricle. This developmental event, which will eventually lead to the formation of the Magendie foramen, was

described at the beginning of the 20th century for the first time by Blake (1) and confirmed after few years by Wilson (2). This structure, which is essentially formed by extremely thin ependyma, has been named by most authors *Blake's pouch,* after its initial describer; the same authors identify with the term of *Blake's pouch cyst* (or *persistence - BPC*) the abnormal – or only delayed - development of the very same structure (1-7, 9-11). Others tend to apply the term of *Blake's pouch* directly to the abnormal development of the same structure, when perforation does not occur at all (8). We will adopt in this study the former terminology. If the further development of the posterior fossa is normal, the Blake's pouch grows and eventually perforates, at the latest by the end of the 10th gestational week, forming the midline aperture of the Magendie foramen (16). It seems that, roughly at the same time, the cisterna magna, which represents a sub-arachnoid space, develops by cavitation from the primitive meninx. As already mentioned, Luschka foramina open much later at 14-17 weeks (12-15) or even later (around 26 weeks – ref.16); it has been also reported that in 1/5th of the cases the lateral recesses of the 4th ventricle may fail to perforate.

b. A**bnormal posterior membranous area (PMA) development: the Blake's pouch cyst/persistence (1-11).** If the *Blake's pouch* does not perforate, it will enlarge to become a cyst-like structure (*Blake's pouch cyst/persistence - BPC*) protruding into the cisterna magna and acting as a wedge for the developing cerebellar vermis, which is located just cranial to the unruptured *Blake's pouch*. As a result, the vermis is passively rotated counterclockwise due to the increasing volume of the unruptured *Blake's pouch*, which has in the meantime become a cyst *(BPC).* In this condition, the vermis itself will be

unremarkable, since the *BPC* derives from the PMA and not from the AMA, as confirmed by the fact that its walls contain ependymal cells but not neurons (8). The IV ventricular choroid plexus will be displaced within the initial tract of the upper wall of the cyst. The cyst itself will be located in the cisterna magna but it is an expansion of the neuraxis (4th ventricle) not in contact with the sub-arachnoid space represented by the cisterna magna. The two terms of *BPC* and *Blake's pouch persistence* have both been used to identify this condition and are synonymous.

2. **Ultrasound and clinical data.** Nineteen cases of BPC represent the study population. Median gestational age at diagnosis was 22 weeks, with only two cases referred in the 3rd trimester (Table 1). Referral indications were: suspect of vermian abnormality in 11 cases (58%), other non-CNS anomaly in the remaining 42% of cases (8/19), with 4 of them being referred for fetal echocardiography due to suspicion of congenital heart disease. As for the ultrasound features of BPC, on the median view of the posterior fossa the upper wall of the cyst was clearly visible in 11/19 cases (fig.1E-L); on the coronal view, choroid plexus tufts were evident in most cases on the supero-lateral borders of the cyst roof (fig.2). The axial trans-cerebellar view was abnormal in all cases, showing the classic keyhole sign (fig.2C). Multiplanar three-dimensional ultrasound was used to enhance the assessment of the BPC and its relationships with the vermis and the posterior fossa (fig.2). Three-dimensional ultrasound with surface-rendering allowed demonstration of the BPC outer wall expanding into the cisterna magna only in 3 cases, due to shadowing from the occipital bone in the majority (fig.3). In most cases, it was noted that the echogenicity of the fluid content of the cisterna magna was hypoechoic, with tiny strands, and not completely translucent as it was within the Blake's cyst (fig.4).

On ultrasound follow up, the BPC disappeared, with the vermis returning to its normal position, in 6 of the 9 cases reaching term of gestation (excluding the case #1 of Table 1, delivered at 28 weeks due to abruption placentae), due to likely late fenestration (fig.5); in the 5/6 cases in which the pregnancies had been followed up at our center, this event took place between 24 and 26 weeks of gestation; in the remaining case, we could only gather postnatal information (MRI) after initial diagnosis at 23 weeks (#16 in Table 1). In case #6, the neonate (dichorionic twin) died soon after birth, and at time of demise, the BPC was still in place, on transfontanellar ultrasound. In the other 3 cases (#4, 10, 15 of Table 1) the moderate counterclockwise vermis rotation persisted until neonatal follow up.

MRI was performed in 15/19 cases at diagnosis of follow up, but in no case did it add anything to the ultrasound diagnosis; in the 5 cases in which it was performed on follow up, after sonographic evidence of late fenestration of the BPC, it confirmed the normal position and anatomy of the vermis and the absence of cysts in the posterior fossa.

Associated anomalies were present in 8 cases (42%) and in 5 of these consisted of or included congenital heart disease (Table 1). Karyotype was normal in 12 cases (5 amniocenteses and 7 postnatal) and abnormal in 2 (trisomy 21); it was not performed in the remaining 5 cases undergoing termination of pregnancy.

As for pregnancy outcome (Table 1), there were 8 terminations of pregnancy (42%), 2 neonatal deaths (10%), and the remaining 9 neonates are alive (48%). One neonate (#16 of Table 1) has obstructive hydrocephaly confirmed (the BPC has disappeared) and is currently being followed up closely; another neonate (#13 of Table 1) was diagnosed with Down syndrome after birth. Neurodevelopmental outcome is normal in 7/8 cases (with the exception of the Down syndrome baby), according to the paediatricians following them, at a mean follow up time of 15 months (range 1-42 months).

DISCUSSION

From the initial description of BPC by Blake in 1900 (1) the publications on BPC have been scant both the neonatal (3-8) and the prenatal literature (9-11). Since no diagnostic criteria have been set so far for diagnosing a BPC in the fetus, we propose that for a BPC to be diagnosed in a fetus the following three criteria should be fulfilled: 1) normal anatomy (including normal appearing fastigium) and size of the vermis; 2) mild to moderate counterclockwise rotation of the vermis; 3) normal size of the cisterna magna. To these, inconstantly, visualization of the BPC roof within the cisterna magna (fig.1-3) and a more translucent echogenicity of the cyst content vs the cisterna magna fluid (fig.4) can also be considered. All of them are visible on the median view of the posterior fossa, whereas the normal size of the cisterna magna can be confirmed also on the axial view. These criteria, a slightly expanded version of those proposed by Malinger (10), would allow in most cases a differential diagnosis vs Dandy-Walker Malformation (DVM), Inferior Vermian Hypoplasia (IVH) and MegaCisterna Magna (MCM) (3, 19-22). *DVM* represents an abnormal development of the AMA characterized by (fig.1D) moderately severe/severe hypoplasia and counterclockwise rotation of the vermis, an elevated insertion of the tentorium, and a large fluid collection in the cisterna magna communicating with the 4th ventricle. *IVH* is also considered an abnormal development of the AMA and is characterized by (fig.1C): moderate to severe hypoplasia of the inferior vermian portion, moderate/moderately severe counterclockwise rotation of the vermis, moderate increase of the fluid collection in the posterior fossa communicating with the 4th ventricle. This represents the most challenging differential diagnosis for BPC, because this is based only on the normal vs abnormal aspect/size of the inferior vermis, considering that BPC and IVH share the moderate counterclockwise rotation of the vermis and the moderate increase of the fluid collection in

the posterior fossa. Furthermore, it is common occurrence to observe a cystic expansion of the 4th ventricle in the cisterna magna also in some cases of IVH, which contributes to the diagnostic challenge. In our experience, also prenatal MRI cannot solve the dilemma in most cases, especially if performed at 20-23 weeks of gestation. *MCM* (fig.1B) represents, similarly to BPC, an abnormality of the PMA and is characterized by a wide collection of fluid in the posterior fossa freely communicating with the arachnoid space and the 4th ventricle, with a vermis of normal size and showing no rotation.

An interesting notation is that the BPC fluid content shows a more translucent echogenicity than that of the cisterna magna (fig.4). Considering that the cisterna magna seems to form from cavitation of the meninges and that the Luschka apertures are still absent, it can be speculated that the differential echogenicity may reflect the different origin of the fluid content: completely anechoic for the BPC, because it consists of cerebrospinal fluid produced from the choroid plexuses; less translucent for the cisterna magna, because this structure would represent only an hollow space deriving from cavitation which will eventually fill with the cerebrospinal fluid once the Luschka apertures and the Magendie foramen will become patent. The fact that in normal conditions the fluid content of the cisterna magna is completely anechoic, with the exception of the thin strands of tissue thought to represent remnants of the Blake's pouch (11), may represent a confirmation of this hypothesis.

Another interesting consideration regards the timing of the opening of Luschka apertures: according to some authors (12-15), this event occurs around 14-17 weeks. However, according to Brocklehurst (16) they become patent around 26 weeks, which is the exact timing of the disappearance of the BPC in the 6 cases of our series in which this event has occurred (Table 1). If this theory is correct, then it may be speculated that an explanation for the disappearance of the BPC alternative to its late fenestration might be deflation, due to

the fact that the opening of the Luschka apertures would null the formerly elevated intra-ventricular pressure, with consequent disappearance of the ballooning. However, if this is the case, then it means that the Magendie foramen might be still closed in those cases in which the BPC has disappeared at 26 weeks.

Moving to the analysis of the natural history of BPC, from our limited data it seems that in roughly half of the cases (6/10) the cyst eventually fenestrates (or disappears), and that, if this event occurs, it occurs at 24-26 weeks of gestation (Table 1). As for the cases in which BPC does not perforate by the time of birth, their longterm outcome will probably depend on the patency and size of the Luschka apertures. At this regard, Tortori-Donati et al. (3) claim that in individuals with BPC there is always a tetraventricular hydrocephaly, due to absence of the Magendie foramen. However, it may be speculated that in some cases the presence of the Luschka apertures ensures a precarious steady state which may become symptomatic only later in life (7) - leading to hydrocephaly - or remain asymptomatic (1, 12).

Another important issue to discuss is the relatively high association rate with major anomalies. In our series, 11/19 cases were isolated and of the 8 associated with other anomalies 5 included a heart defect (Table 1). In addition, 10% (2/19) of all cases and 14% (2/14) of those with a known karyotype had Down syndrome. The series is of course too limited to draw any conclusion on whether the recognition of a BPC should prompt fetal echocardiography or karyotyping. However, in this case the usual bias of fetal series – the fact that cases associated with major abnormalities are preferentially referred for expert opinion – does not hold. In fact, >50% of the cases were isolated and these were indeed referred because the trans-cerebellar view, which is part of the 2nd trimester anomaly scan checklist (17, 23), was abnormal, showing the classic *keyhole sign* (fig.2C).

The final comment regards the neurological outcome. BPC derives from the PMA and, as such, does not contain neurons (8). This very fact is the reason why neurodevelopmental outcome is generally normal in neonates and infants with a BPC (3, 4-6). This concept is supported also by our data: the outcome was normal in all 9 cases with isolated BPC that survived the early neonatal period. Although, admittedly, the normal neurological outcome was only ascertained by contacting the family paediatricians who followed them, nonetheless a major neurodevelopmental would certainly be evident.

Another important limitation of the study is that it is very difficult to confirm the presence of a BPC at necropsy. In most cases undergoing termination of pregnancy, what could be confirmed without doubt was that the anatomy of the cerebellar vermis was normal, because dissection of the posterior fossa caused the BPC to empty and the vermis to return to its normal position. in these circumstances, a post-mortem MRI may help.

In conclusion, we have described the pathogenesis of the BPC, collating evidence from different embryological (1,2, 12-16) and pediatric neuroradiological (3-7) studies. In addition, we have proposed the criteria to diagnose a BPC in the fetus, showing also that the cyst may undergo delayed fenestration at 24-26 weeks of gestation. Finally, we have reported on the apparent significant risk of associated anomalies, including CHD and trisomy 21. Further fetal series are needed to confirm the concepts expressed in this study. However, it should be considered that this, to the best of our knowledge, represents the first analysis of BPC in the fetus, from diagnosis to natural history, associations and outcome.

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FIGURE LEGENDS

1. Cystic anomalies of the posterior fossa, as evident on median view of the fetal head, approached transvaginaly from the posterior fontanelle. A) normal aspect at 22 weeks of gestation: the vermis is very close to the pons; the fastigium, the 4th ventricle and the cisterna magna are clearly visible; B) Megacisterna Magna at 32 weeks of gestation. In this case, the fluid collection in the posterior fossa is rather large, displacing also the tentorium (usually this finding is not seen at midtrimester); the position of the vermis is similar to that in A. C) Inferior Vermian Hypoplasia (IVH) at 21 weeks of gestation. The fastigium cannot be recognised, the lowermost portion of the vermis is absent, the vermis itself show pronounced counterclockwise rotation; D) Dandy-Walker Malformation (DWM) at 28 weeks of gestation. The tiny hyperechoic vermis is barely visible and is surrounded by a large fluid cystic collection in the posterior fossa; the insertion of the tentorium is displaced cephalad. E-L: Blake's Pouch Cyst (BPC) during the 2nd (E-G, I-K) or the 3rd trimester of pregnancy (H and L). Note the normal aspect of the vermis, with normal folia, and a variable degree of anti-clockwise rotation (maximum in F and G, minimal in K). In all, the vermis appears detached from the pons (compare with A). **In reference to Table 1, the correspondence is as follows: E-15, F-11, G-10, H-4, I-13, J-5, K-9, J-6.**
2. Blake's Pouch Cyst at 22 weeks of gestation. Three-dimensional ultrasound with multiplanar and volume contrast imaging. A) on the median view, the anti-clockwise rotation of the normal vermis (V) and the roof of the Blake's pouch cyst (arrowhead) are evident; B) on the corresponding coronal view, two tufts of choroid plexus (arrowhead) can be seen on the supero-lateral margins of the cyst wall, just below the vermis (V); C) on the axial plane, the classic "keyhole sign" (arrow) is seen, with non-visualization of the vermis in between the two cerebellar hemispheres. This sign

leads to referral for expert opinion and final diagnosis. The lateral walls of the cyst are evident (arrowheads).

3. Three-dimensional ultrasound with surface rendering. A) to render the outer surface of the BPC, the Region Of Interest (ROI) box should be in the posterior fossa, with the green bar slightly curved on the upper surface of the cerebellum. In this way, it is possible to demonstrate the digit-like expansion of the cyst protruding into the cisterna magna (arrows), just below the cerebellar vermis (V). The arrowheads identify the occipital bone; B) to confirm the finding, a normal posterior fossa is shown here. the same technique shows only the area corresponding to the empty cisterna magna (CM), with no evidence of any cystic structure below the vermis (V).
4. Three-dimensional ultrasound with multiplanar and volume contrast imaging. Using a high frequency (6-12MHz) transvaginal transducer, the fluid content of the Blake's pouch (BPC) appears completely sonolucent, whereas the content of the cisterna magna (arrows) shows some grainy textures with thin vertical strands. A) median view; B) coronal view. (OH: occipital horns of the lateral ventricles)
5. BPC can undergo delayed fenestration (A-B) or persist until birth (C-D). Median view. A) BPC at 20 weeks of gestation: the classic aspect of BPC is seen, with an anatomically normal vermis (V) showing anti-clockwise rotation. The BPC (asterisk) and its roof (arrow) are visible; B) the same case at 28 weeks of gestation: the vermis (V) in no longer rotated counterclockwise, and has returned to its normal position close to the pons. A normal 4th ventricle is visible (it was not before), and there is not any sign anymore of the former BPC. C) another case of BPC detected at 22 weeks of gestation. For description and captions see A; D) at 33 weeks of gestation, the situation is the same, with an counterclockwise rotated vermis (V), the BPC (asterisk),

with its roof (arrow). The same image was present also on postnatal MRI pwrformed at 1 month of age.

Table 1. Characteristics, associations and outcome of the 19 cases of Blake's pouch cyst.

Case Number	Indication to ultrasound	GA (wks)	Associated anomalies	Karyotype	Fenestration (GA in wks)	Outcome (follow up)
1	Vermian abn.?	24	No	Normal	No[1]	NND[1]
2	Vermian abn.?	22	NF	Normal	--	TOP
3	ECA	18	Bilateral Multikystic Kidney	NA	--	TOP
4	Vermian abn.?	34	No	Normal[2]	No	AW (42 months)
5	Vermian abn.?	20	No	Normal	Yes (26)	AW (36 months)
6	CHD	30	TOF + LSVC + agenesis DV	NA	No	NND[3]
7	Vermian abn.?	21	No	Normal[2]	Yes (24)	AW (24 months)
8	CHD	23	DISV + aortic arch abnormality	Normal	--	TOP
9	Vermian abn.?	24	No	Normal[2]	Yes (24)	AW (21 months)
10	Vermian abn.?	23	No	Normal[2]	No	AW (10 months)
11	CHD	22	TOF	NA	--	TOP
12	Vermian abn.?	23	No	NA	--	TOP
13	Vermian abn.?	24	No	Trisomy 21[2]	Yes (26)	Alive (6 months)
14	CHD	22	PA+VSD + pACC	Normal	--	TOP
15	Vermian abn.?	20	No (IUGR)	Normal[2]	No	AW (1 month)
16	ECA	23	Ventriculomegaly	Normal[2]	Yes (NA)	Alive (2 months)
17	Vermian abn.?	22	No	Normal[2]	Yes (25)	AW (1 month)
18	ECA	23	Clubfeet + Micrognathia	NA	--	TOP
19	ECA	19	pAVSD + NF + Absent NB	Trisomy 21	--	TOP

Abn.: abnormality; AW: alive and well; CHD: congenital heart disease; DISV: double inlet single ventricle; DV: ductus venosus; ECA: extra-cardiac anomaly; IUGR: intra-uterine growth retardation; LSVC: left superior vena cava; NA: not available; NB: nasal bone; NF: nuchal fold > 5 mm; TOF: tetralogy of Fallot;

[1]: preterm delivery from abruptio placentae at 28 weeks. Severe cerebral haemorrhage

[2]: on postnatal examination

[3]: dichorionic twin pregnancy; the co-twin is alive and well. The affected neonate weighed 1090 grams at birth (38 weeks) and died soon after due to severe IUGR and multiple anomalies.

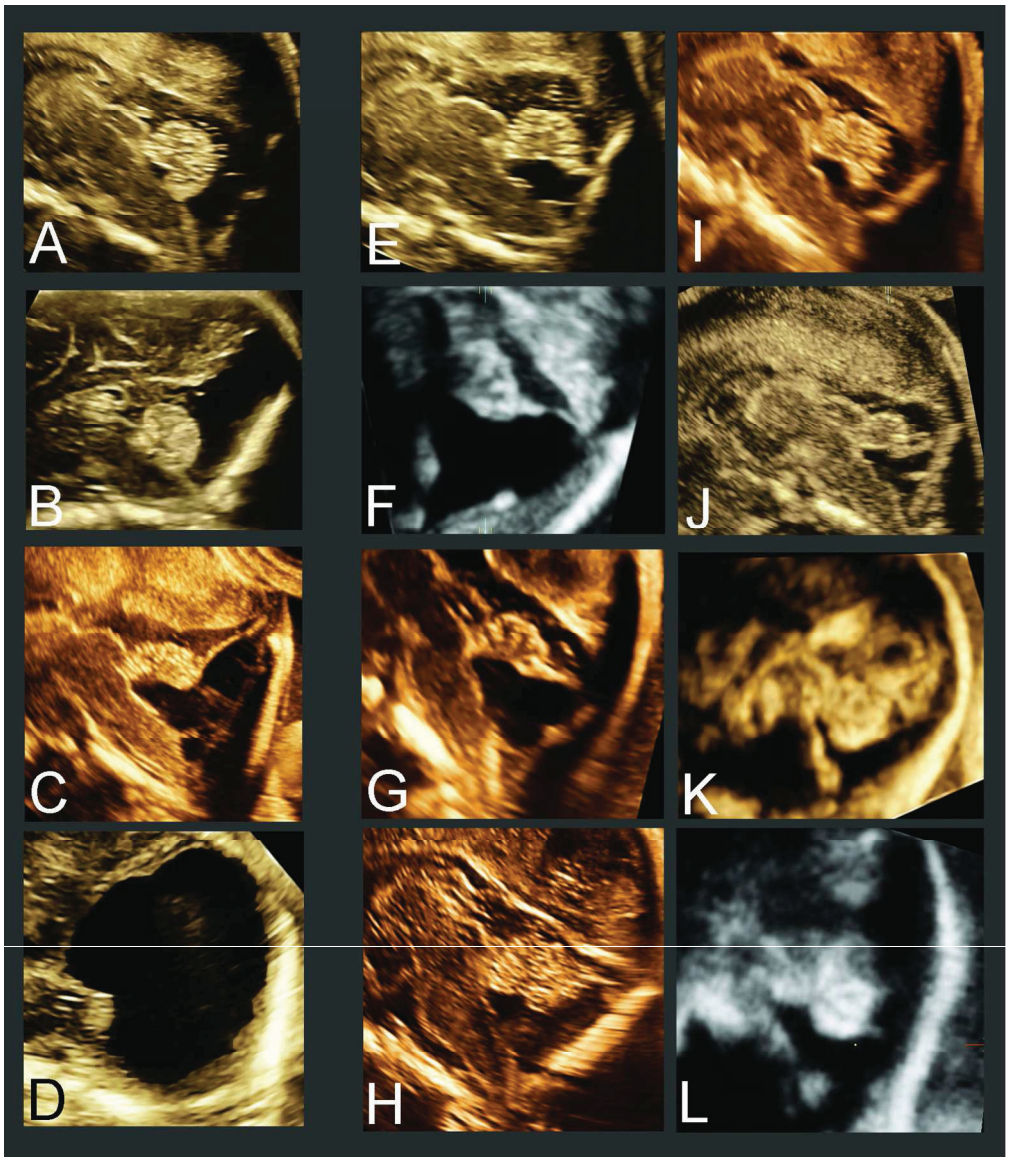

fig 1. Cystic anomalies of the posterior fossa, as evident on midsagittal view of the fetal head, approached transvaginaly from the posterior fontanelle. A) normal aspect at 22 weeks of gestation: the vermis is very close to the pons; the fastigium, the IV ventricle and the cisterna magna are clearly visible; B) Megacisterna Magna at 32 weeks of gestation. In this case, the fluid collection in the posterior fossa is rather large, displacing also the tentorium (usually this finding is not seen at midtrimester); the position of the vermis is similar to that in A. C) Inferior Vermian Hypoplasia (IVH) at 21 weeks of gestation. The fastigium cannot be recognised, the lowermost portion of the vermis is absent, the vermis itself show pronounced anticlockwise rotation; D) Dandy-Walker Malformation (DWM) at 28 weeks of gestation. The tiny hyperechoic vermis is barely visible and is surrounded by a large fluid cystic collection in the posterior fossa; the insertion of the tentorium is displaced cephalad. E-L: Blake's Pouch Cyst (BPC) during the 2nd (E-G, I-K) or the 3rd trimester of pregnancy (H and L). Note the normal aspect of the vermis, with normal folia, and a variable degree of anti-clockwise rotation (maximum in F and G, minimal in K). In all, the vermis appears detached from the pons (compare with A).

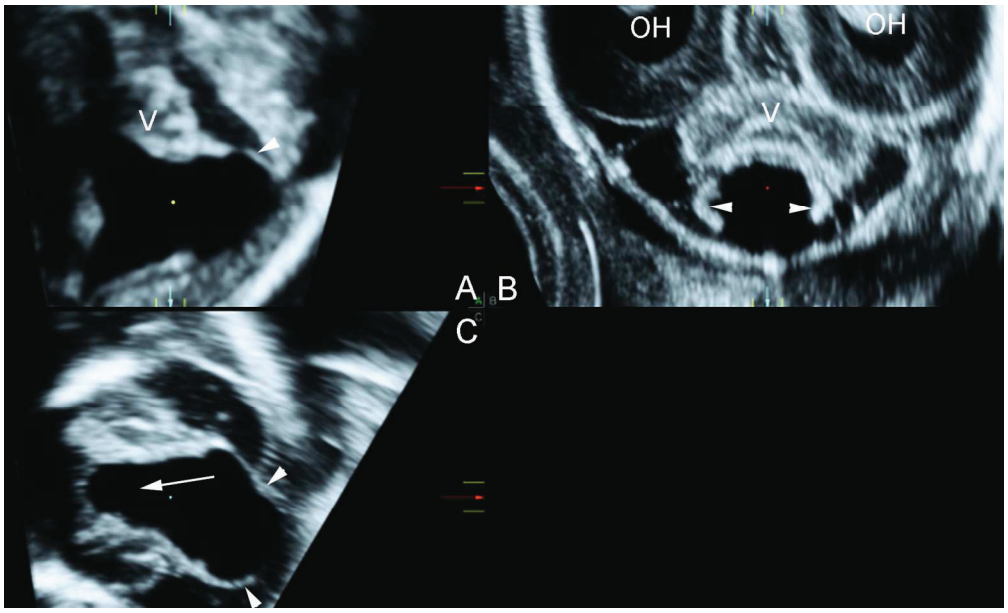

Fig. 2. Blake's Pouch Cyst at 22 weeks of gestation. Three-dimensional ultrasound with multiplanar and volume contrast imaging. A) on the midsagittal view, the anti-clockwise rotation of the normal vermis (V) and the roof of the Blake's pouch cyst (arrowhead) are evident; B) on the corresponding coronal view, two tufts of choroid plexus (arrowhead) can be seen on the supero-lateral margins of the cyst wall, just below the vermis (V); C) on the axial plane, the classic "keyhole sign" (arrow) is seen, with non-visualization of the vermis in between the two cerebellar hemispheres. This sign leads to referral for expert opinion and final diagnosis. The lateral walls of the cyst are evident (arrowheads).

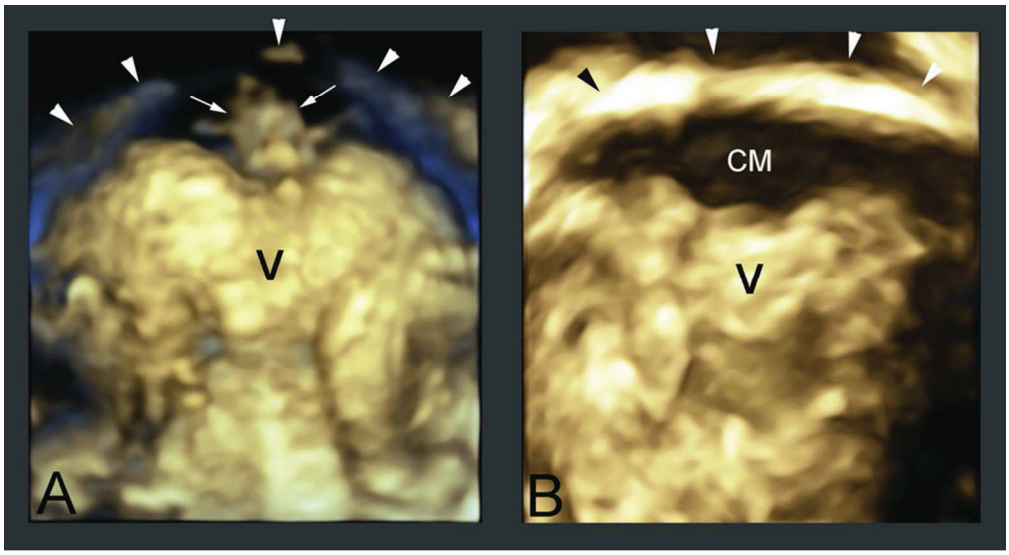

Fig.3. Three-dimensional ultrasound with surface rendering. A) to render the outer surface of the BPC, the Region Of Interest (ROI) box should be in the posterior fossa, with the green bar slightly curved on the upper surface of the cerebellum. In this way, it is possible to demonstrate the digit-like expansion of the cyst protruding into the cisterna magna (arrows), just below the cerebellar vermis (V). The arrowheads identify the occipital bone; B) to confirm the finding, a normal posterior fossa is shown here. the same technique shows only the area corresponding to the empty cisterna magna (CM), with no evidence of any cystic structure below the vermis (V).

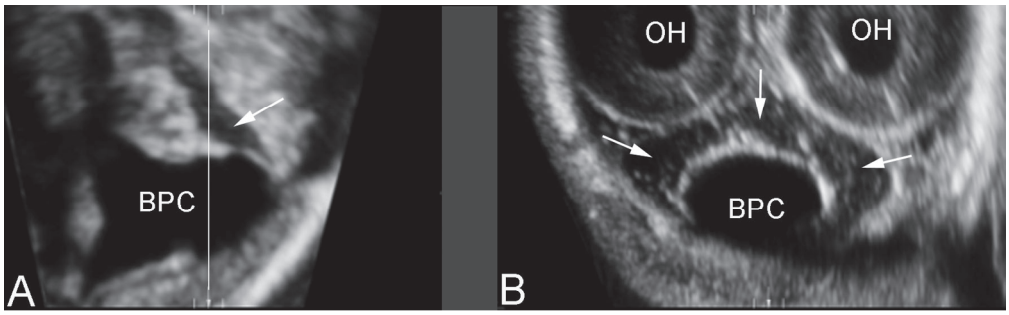

Fig.4. Three-dimensional ultrasound with multiplanar and volume contrast imaging. Using a high frequency (6-12MHz) transvaginal transducer, the fluid content of the Blake's pouch (BPC) appears completely sonolucent, whereas the content of the cisterna magna (arrows) shows some grainy textures with thin vertical strands. A) midsagittal view; B) coronal view. (OH: occipital horns of the lateral ventricles)

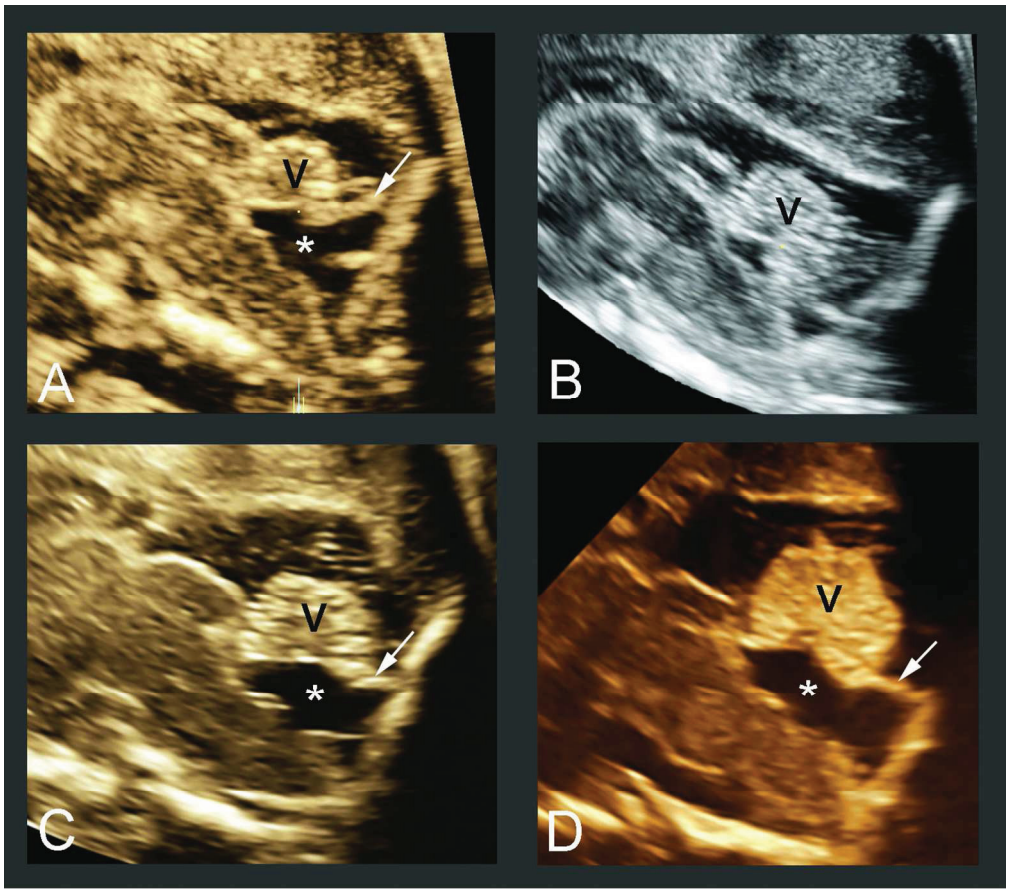

Fig.5. BPC can undergo delayed fenestration (A-B) or persist until birth (C-D). Midsagittal view. A) BPC at 20 weeks of gestation: the classic aspect of BPC is seen, with an anatomically normal vermis (V) showing anti-clockwise rotation. The BPC (asterisk) and its roof (arrow) are visible; B) the same case at 28 weeks of gestation: the vermis (V) in no longer rotated anticlockwise, and has returned to its normal position close to the pons. A normal IV ventricle is visible (it was not before), and there is not any sign anymore of the former BPC. C) another case of BPC detected at 22 weeks of gestation. For description and captions see A; D) at 33 weeks of gestation, the situation is the same, with an anticlockwise rotated vermis (V), the BPC (asterisk), with its roof (arrow). The same image was present also on postnatal MRI pwrformed at 1 month of age.