The Cerebellar Vermis Made Simple

Ilan E. Timor-Tritsch & Ana Monteagudo
Disclosure

• No issues to disclose
Objectives

• The participants will be able to:
  • Refresh their long forgotten embryology and anatomy of the cerebellum
  • Be able to differentiate between the subtle changes of vermian position from normal to pathological
  • Judge if there is a need an MRI for added diagnostic workup
Introduction

• It is impossible to speak about the vermis without speaking about the cerebellar hemispheres & cisterna magna.

• That, since many pathologies that affect the vermis also disrupt the normal appearance of the cerebellum and cisterna magna.
What does the vermis do?

- Proprioception
  - ability to recognize the relative positioning of body parts used for movement
- Control of muscle tone and level of force.
• AIUM guidelines include imaging of the cerebellum, cisterna magna, and lateral ventricles; as part of the minimal elements of the fetal anatomy scan.

• When normal, it has a very high negative predictive value

NPV is the probability that fetuses with a negative screening test truly don’t have the disease.
Cerebellum: Quick Facts

• Transcerebellar diameter (TCD)
  – Doubles in size during 2\textsuperscript{nd} trimester
  – Between 18-24 wks the TCD = number of weeks ± 1mm
    • Useful in cases of IUGR / macrosomia
    • Dating, Brain sparing?

Cisterna Magna: Quick Facts

(“anatomically” correct term: cerebellomedullary cistern)

- Size in the 2\textsuperscript{nd} trimester is stable
- Mean size: 5 ± 3mm
- Upper limit of normal: 10 mm

Mahoney B et al. Radiology 1984;153:773
Cisterna magna septa: Quick Facts

- Seen in 84-92% of fetuses (2\textsuperscript{nd} 3\textsuperscript{rd} ∆)
- Usually 2 septae are seen
- Found inferior & posterior from the vermis
- Straight & parallel with the AP diameter
- These are NL anatomic structures a result of NL embryologic development of the posterior fossa**
  ** Represent the walls of Blake’s pouch

* Pretorius DH et al, JUM1992;11:125; * Knutzon RK et al, Radiology 1991;190:70
  ** Robinson AJ & Goldstein R, JUM2007;26:83
Blake’s Pouch a.k.a. Cisterna Magna Septa

Quick Facts

• The cisterna magna septa are the walls of Blake’s pouch.
• Blake’s pouch is a normal fingerlike appendage of the 4th ventricle.
• ‘Potential marker’ for normal development.

Pretorius DH et al, JUM1992;11:125
Knutson RK et al, Radiology 1991;190:70
Robinson AJ & Goldstein R, JUM2007;26:83
Sonographic Appearance
Sidewalls of Blake’s pouch
Anechoic fluid
Slightly low-level echoic fluid in CM (subarachnoid space)
Cerebellum
Blake’s Pouch
Early Vermian Development

• When scanning in the axial plane a connection between the 4\textsuperscript{th} ventricle and cisterna magna may be seen between 15-18 weeks

• Question:
  – Is this a malformation? Is this vermian hypoplasia (Dandy-Walker variant)?
Early Vermian Development

Answer:

No- this gap is known as Blake’s metapore as it contains the neck of Blake’s pouch. Once Blake’s pouch fenestrates, the metapore is known as the foramen of Magendie.

Typically seen when the “axial section” is not perfect and the section is semi-coronal.

To avoid making the error and having the right plane, ensure that cavum septi pellucidi is seen in the axial section.
Walls of Blake pouch cyst

Fetus < 20 weeks - seen in semicoronal plane.

Walls of Blake pouch cyst

Blake' metapore

Subarachnoid space

Brainstem

Blake pouch cyst
Which of the 3 scanning planes provides the most information about the vermis?
Transcerebellar Plane

Q: Is this the right plane?

- Vermis is located between the 2 cerebellar hemispheres
- However, in this plane it is difficult to adequately image and evaluate the vermis. Since its borders are not well demarcated
The occipital horns are seen
The tentorium is seen
Vermis is located between the 2 cerebellar hemispheres
However, the borders of the vermis are not well demarcated
Median plane

Q: Is this the right plane?

YES ... it is!!
• The median plane allows easy visualization of the vermis
• The vermis is found in the midline between 2 the cerebellar hemispheres
Median Section: ‘The Posterior Fossa’

- Part of intracranial cavity
- Located between the tentorium cerebelli & foramen magnum
- Houses:
  - Cerebellum
  - Vermis, 4V
  - Cisterna magna septa
  - Brainstem
  - Pons and medulla
Tentorium Cerebelli

- 2nd largest dural fold
- Attached to falx cerebri at its midline
- It separates the cerebellum from the occipital lobes of the brain.
- It encloses the transverse sinus
Tentorium Cerebelli
**Torcular Herophili**  
a.k.a **Confluence of the sinuses**

- It is the point where the superior sagittal sinus, straight sinus and occipital sinus meet.
Tentorium Cerebelli

Confluence of the sinuses or Torcular herophili
Superior sagittal sinus

Straight sinus

Median Plane
Confluence of the sinuses or
Torcular Herophili
Why is the Tentorium & Torcular Herophili important??

Because: Its elevation is a sensitive indicator of posterior fossa pathology.

Confluence of the sinuses or Torcular
The Vermis
The median plane

- Is the best plane to evaluate the vermis
- Echogenic
- Vermis ‘sits’ on the brainstem
• Primary Fissure
  • Separates the anterior from the posterior lobe

• Anterior lobe
  • Lingula, central lobule, culmen

• Posterior lobe
  • Horizontal, prepyramidal and secondary fissures

Ratio of 1:2 between the superior and inferior part of the vermis
The Vermis- Primary Fissure
The median plane

- The 1st vermian fissure is deep and separates the anterior lobe from the posterior lobe.
- Its identification suggests normal development of the vermis; although it does not eliminate other conditions.
- Consistently seen ≥ 24 weeks.

Early Vermian Development

FIGURE 1. Diagrammatic representation of early vermian development as seen by MR imaging of fetal specimens. ¹⁹–²¹ Eleven to 12 weeks—developing vermis (A, single arrow) and position of fourth ventricle roof which is nonresoluble by MR (double arrow). Thirteen to 14 weeks—developing fastigial point (B, single arrow) and fourth ventricle roof now visible. The brain stem develops a kink—the dorsal pontine flexure (double arrow). Sixteen weeks—vermis now covers fourth ventricle roof (C, double arrow). Eighteen weeks—vermis usually complete (D, double arrow).
Early Vermian Development
Blake’s Pouch

Figure 1 (a) During formation of the dorsal pontine flexure (small arrow) a transverse crease (large arrow) forms in the roof of the rhombencephalic vesicle (*), dividing it into anterior (cranial) and posterior (caudal) membranous areas. (b) The vermis (arrowhead) develops from the rhombic lip at the superior margin of the anterior membranous area. Choroid plexus develops in the crease (arrow). Cavitation starts in the overlying meninx primitiva (double arrow) to form the subarachnoid space. (c) As the cerebellum grows inferiorly the posterior membranous area bulges out between the vermis (large arrow) and the nucleus gracilis (small arrow), forming Blake’s pouch. The subarachnoid space remains trabeculated by pia-arachnoid septations (double arrow). (d) Blake’s pouch fenestrates (dotted line) and the neck of Blake’s pouch becomes the foramen of Magendie (dashed line). The choroid plexus (arrow) now appears to be in the cisterna magna.

(All images reproduced, with permission of the American Institute of Ultrasound in Medicine, from Robinson and Goldstein15.)

Blake’s pouch normal embryological structure. Failure of the normal fenestration to occur; results in persistent Blake’s Pouch (Cyst)
Definitions

- Blake’s pouch cyst (Persistent Blake’s pouch) - Apparent communication between 4\textsuperscript{th} ventricle and the posterior fossa with a normal vermis, the vermian fissures and fastigium are also normal.

• Dandy-Walker malformation
  – Complete or partial vermian agenesis, cystic dilatation of the 4th ventricle (persistent BPC) with enlarged posterior fossa, rotation of vermis and elevation of the tentorium and torcula.

Definitions

• Vermian abnormalities (a.k.a DWM variant)
  – Agenesis (absent), hypoplastic (present but small)

Editorial

Inferior vermic hypoplasia – preconception, misconception

ASHLEY J. ROBINSON
Department of Radiology, Children’s Hospit.
Columbia, 4480 Oak Street, Vancouver V6H (e-mail: ash@radiologist.net)
Vancouver, 4480 Oak Street, Vancouver V6H (e-mail: ash@radiologist.net)

Embryology of the posterior fossa and the hindbrain

The neural tube

Introduction

There is considerable confusion in the literature regarding the terminology used when describing abnormalities of the cerebellum and of the vermis in particular. Terminology such as ‘closure of the fourth ventricle’, ‘craniocaudal growth of the vermis’ and ‘inferior vermic hypoplasia’, as well as the numbering (using Roman numerals) of the cerebellar lobules from anterior to posterior, has left us with the preconception and misconception that the vermis grows from superior to inferior, and that partial agenesis or hypoplasia always involves the inferior lobules. In light of recent advances in our understanding of the embryology of the cerebellum and cisterna magna, certain terminology and concepts can be demonstrated to be incorrect and should be abandoned.
The Vermis: Upward rotation

- Can be associated with benign asymptomatic conditions to severe brain abnormalities with significant neurological impairment.
- Example: Blake’s pouch cyst, vermian hypoplasia and DWM
- How can this rotation be quantified?
Brainstem–vermis and brainstem–tentorium angles allow accurate categorization of fetal upward rotation of cerebellar vermis.

1) Brainstem-vermis angle (BV)- a line is drawn tangentially to the dorsal aspect of the brain stem. A second line is drawn tangentially to the ventral contour of the vermis.

2) Brainstem-tentorium angle- a third line is drawn tangentially to the tentorium.
Application of BV and BT angles in fetuses with posterior fossa abnormality

**Blakes’ pouch cyst**

In Blakes’ pouch:
- BV: 23 ± 2.8° (19-26°)
- BT: 42.2 ± 7.1° (32-52°)

**Cerebellar vermis hypoplasia**

In vermian hypoplasia:
- BV: 34.9 ± 5.4° (24-40°)
- BT: 52.1 ± 7.° (45-66°)

**Dandy-Walker malf.**

In Dandy-Walker malf:
- BV: 63.5±17.6° (45-112°)
- BT: 67.2 ± 15.1° (51-112°)
Conclusions

- BV angle < 18° and BT angle < 45° was always found in the normal controls
- BV angle > 45° - is strongly suggestive of DWM
- BV angle < 30° suggests a dx. Blake’s pouch cyst
- BV angle increases with increasing severity of the condition
- BT angle showed similar pattern; but there was much overlap between the groups
- BV angle objective finding useful in differentiating posterior fossa fluid collections
• When scanning in the median plane a connection between the 4th ventricle and cisterna magna may be seen between 15-18 weeks
• Question:
  – Is this normal? Is it persistent Blake’s pouch cyst? Is this a Dandy-Walker?
Objective: To evaluate the role of the brainstem–vermis (BV) angle in the diagnosis of fetal posterior fossa abnormalities at 15–18 weeks’ gestation.

Methods: Retrospective review of 3D volumes. Whether the fourth ventricle appeared open posteriorly in axial views was noted and the BV angle was measured.
Normal fetus with ‘closed’ 4th ventricle
BV angle < 20°

Normal fetus with ‘open’ 4th ventricle; similar appearance to Blake’s Pouch Cyst
BV angle 20° - 40°

Fetus with Dandy-Walker malformation
BV angle > 45°

**Conclusion:** An open 4th ventricle is found in about 10% of normal fetuses at 15–18 wks’. Measurement of the BV angle is useful in these cases, as a value ≥45° is associated with a very high risk of severe posterior fossa malformation.
Answer

- Connection between the 4\textsuperscript{th} ventricle and cisterna magna between 15-18 weeks is seen in approximately 10\% of normal fetuses
  - BV angle < 20° is ‘normal’
- In cases diagnosed with Blake’s pouch cyst the BV angle was between 20-40°
- In cases with the Dandy-Walker malformation the BV angle was > 45°
The Vermis: Is it too small

Table 2—Imaging findings in patients with congenital cerebellar malformations

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>TCD</th>
<th>Vermis biometry</th>
<th>Sup/Inf ratio</th>
<th>Fastigium</th>
<th>Fourth ventricle</th>
<th>Pons</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delayed ‘closure’</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Open</td>
<td>N</td>
<td>Good</td>
</tr>
<tr>
<td>Vermian hypoplasia</td>
<td>N</td>
<td>Small</td>
<td>N</td>
<td>N</td>
<td>Open/closed</td>
<td>N</td>
<td>Variable</td>
</tr>
<tr>
<td>Vermian agenesis</td>
<td>N/S</td>
<td>Small</td>
<td>Abn</td>
<td>Abn</td>
<td>Abn</td>
<td>N</td>
<td>Malformations?</td>
</tr>
<tr>
<td>DWM</td>
<td>N</td>
<td>—/Small</td>
<td>Abn</td>
<td>Abn</td>
<td>Abn</td>
<td>N</td>
<td>Poor</td>
</tr>
<tr>
<td>MTRS</td>
<td>N/S</td>
<td>—/Small</td>
<td>Abn</td>
<td>Abn</td>
<td>Abn</td>
<td>?</td>
<td>Poor</td>
</tr>
<tr>
<td>Pontocerebellar hypoplasia</td>
<td>S</td>
<td>Small</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Abn</td>
<td>Poor</td>
</tr>
<tr>
<td>Rhombencephalosynapsis</td>
<td>S</td>
<td>—</td>
<td>—</td>
<td>Abn</td>
<td>Abn</td>
<td>N</td>
<td>Poor</td>
</tr>
</tbody>
</table>

TCD, Transverse cerebellar diameter; Sup/Inf, ratio between the superior and inferior portions of the vermis; N, normal; S, small; Abn, abnormal; Malformations?, depends on the presence of associated malformations; —, agenesis. MTRS: molar tooth related syndromes.

- Vermis
  - Biometry
  - 2 vermian fissures seen, with a ratio of 1 : 2 between the superior and inferior part
  - Fastigium

Malinger G et al. The fetal cerebellum. Pitfalls in diagnosis and management. Prenat Diagn 2009:
Perform all measurements of the *vermis* and the posterior fossa.
Vermis

- 3D acquisition in the axial plane
- Linear relationship

Figure 5. Linear growth of the vermis during pregnancy (18–26 weeks). The solid line is the linear regression line for vermis length; the dotted lines represent ±2 SD of the mean for each gestational week.

Table 2. Measurements of Fetal Vermian Length by Gestational Age

<table>
<thead>
<tr>
<th>Gestational Age, wk</th>
<th>Measurement by Percentile, mm</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>5th</td>
</tr>
<tr>
<td>18 and 19a</td>
<td>8.4</td>
</tr>
<tr>
<td>20</td>
<td>8.8</td>
</tr>
<tr>
<td>21</td>
<td>10.1</td>
</tr>
<tr>
<td>22</td>
<td>11.3</td>
</tr>
<tr>
<td>23</td>
<td>11.8</td>
</tr>
<tr>
<td>24</td>
<td>12.61</td>
</tr>
<tr>
<td>25</td>
<td>13.6</td>
</tr>
<tr>
<td>26</td>
<td>15.1</td>
</tr>
</tbody>
</table>

ND indicates not determined.
aCombined to adjust for small numbers.
Increased Fluid in the Posterior Fossa

- Dandy-Walker cyst
- Blake’s pouch cyst
- Vermian hypoplasia
- Megacisterna magna
Increased Fluid in the Posterior Fossa

Three Important Questions

• Is the vermis rotated?
• Is the torcular elevated?
• Is the vermis normal sized or hypoplastic?
Dandy-Walker Malformation

- Is the vermis rotated?
  - Yes, BV angle increased (140°)

- Is the torcular elevated?
  - Yes

- Is the vermis normal sized or hypoplastic?
  - Yes, hypoplastic
DWM:
- Upward rotation of the vermis (BV = 60°)
- Hypoplastic vermis
- Elevated torcular
Dandy-Walker Malformation

- Prevalence 1:25,000 to 1:30,000 newborns
- Most cases are sporadic
- However, can be cause by gene mutations
- Is associated with chromosomal abnormalities
  - Trisomy (T18, 21, 13 and 9) and triploidy
- Could be caused by environmental factors
  - Infections (rubella, Toxo) and teratogens
- Recurrence risk is low 1-2% for non-syndromic
- Prognosis: intellectual development is variable
Joubert syndrome

Vermian hypoplasia
Molar Tooth sign
Joubert syndrome

- Affect between 1: 80,000 to 1:100,000 newborns
- Autosomal recessive; rarely X-linked recessive
- 10 mutation – these lead to problems with structure and function of cilia (ciliopathy)
- Classic: molar tooth sign (MTS), Hypotonia, Developmental delays
Blake’s Pouch Cyst

- Is the vermis rotated?
  - Yes (BV angle between 20-40°)

- Is the torcular elevated?
  - No

- Is the vermis normal sized or hypoplastic?
  - Yes, normal

Blake’s Pouch Cyst

- Mild rotation
- $BV = 30^\circ$
- Normal vermis.
- Normal torcular
Blake’s Pouch Cyst
Blake’s Pouch Cyst

BV=40°
29\(\frac{1}{7}\) wks Second opinion for a DWM

What’s the diagnosis?

Blake’s Pouch Cyst
3 criteria for diagnosis of BPC:
- 1) normal anatomy and size of the vermis
- 2) mild/moderate rotation of the vermis
- 3) normal size of the cisterna magna.

BPC can undergo delayed fenestration at 24–26 weeks in more than 50% of cases.

BPC shows a risk of association with extracardiac anomalies (heart defects in particular) and, to a lesser extent, T21.
Vermian Hypoplasia

• Is the vermis rotated?
  – No, BV = 30°

• Is the torcular elevated?
  – No

• Is the vermis normal sized or hypoplastic?
  – Yes, hypoplastic

Mega-cisterna Magna

- Is the vermis rotated?
  - No
- Is the torcular elevated?
  - No
- Is the vermis normal sized or hypoplastic?
  - Yes, normal
Table 2 Sonography in fetuses with posterior fossa fluid collections (PFFC), associations with other anomalies, intrauterine regression, outcome and accuracy of prenatal diagnosis

<table>
<thead>
<tr>
<th>Prenatal sonographic diagnosis</th>
<th>Total cases (n)</th>
<th>Cases with associated anomalies (n)</th>
<th>Lost to follow-up (n)</th>
<th>TOP (n)</th>
<th>Regression in utero (n)</th>
<th>Sonographic diagnosis confirmed (n)</th>
<th>Abnormal neurological development postnatally†</th>
<th>Isolated PFFC (n)</th>
<th>PFFC with associated anomalies (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blake’s pouch cyst</td>
<td>32</td>
<td>8</td>
<td>3</td>
<td>2</td>
<td>11/27</td>
<td>16/18</td>
<td>1/20</td>
<td>1/20</td>
<td>1/5</td>
</tr>
<tr>
<td>Megacisterna magna</td>
<td>27</td>
<td>9</td>
<td>4</td>
<td>2</td>
<td>6/21</td>
<td>16/17</td>
<td>2/16</td>
<td>2/16</td>
<td>1/4</td>
</tr>
<tr>
<td>Dandy–Walker malformation</td>
<td>26</td>
<td>16§</td>
<td>7</td>
<td>11</td>
<td>0/8</td>
<td>16/19</td>
<td>3/5</td>
<td>3/5</td>
<td>2/2</td>
</tr>
<tr>
<td>Vermian hypoplasia</td>
<td>17</td>
<td>11§</td>
<td>9</td>
<td>2</td>
<td>0/6</td>
<td>6/8</td>
<td>1/3</td>
<td>1/3</td>
<td>2/2</td>
</tr>
<tr>
<td></td>
<td>102</td>
<td>44</td>
<td></td>
<td></td>
<td>17/72</td>
<td>62/70</td>
<td>7/34</td>
<td>6/13</td>
<td>6/13</td>
</tr>
</tbody>
</table>

*Denominator excludes cases lost to follow-up and terminations of pregnancy (TOP). †Confirmation postnatally or at autopsy; denominator excludes cases lost to follow-up and those that underwent intrauterine regression. ‡Number of cases with abnormal neurological development/number of cases that underwent neurological examination at 1–5 years. §In one case in each group the presence of associated anomalies was detected only postnatally.
In Summary...

• The median section is an indispensable plane when evaluating and diagnosing vermian pathology
In Summary...

Three important questions

- Is the vermis rotated?
- Is the torcular elevated?
- Is the vermis normal sized or hypoplastic?
In Summary...

- The BV angle can help in the differential dx:
  - Normal < 20°
  - Blake’s pouch 20-40°
  - Dandy-Walker > 45°
  - The greater the BV angle the ‘worst’ the diagnosis.
Conclusion

• This lecture is but the tip of the iceberg; when dealing with vermician pathology.

• The vermis is a highly complex structure which we are just beginning to understand.