Fetal Ventricular Dilation; Prelude to Dandy-Walker Syndrome and Hydrocephalus: Synopsis of Two Case Reports

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Abstract

Accurate measurement of the 3rd and 4th ventricles is important in cases of prenatal and postnatal diagnosis of suspected cranial anomalies. Ventricular dilation occurs with excess fluid in the embryologic ventricles during brain development, we make comparative analysis of two case reports on a 23rd week fetus and a 10-month-old neonate. Ultrasound finding related to horns and ventricular system through transfontanelle ultrasound (excluding any > 95th error margin and < 5th percentile) can be utilized for a specific diagnosis of ventricular dilation. Hydrocephalus is defined as accumulation of excessive CSF resulting in dilation of cerebrospinal compartment in the calvaria; which can be acquired or congenital in origin. Hydrocephalus can also be seen due to obstructed flow, faulty absorption and overproduction of CSF (in the subarachnoid space) by a choroid plexus papilloma. Diameter of the anterior horn of the RT and LT (hydrocephalic) lateral ventricles measured 15.3mm and 14.1mm respectively, far above threshold for normal (control) neonates / RT and LT (anterior) lateral ventricles averaging 2.40mm and 2.51mm. These documented findings indicate the excellent agreement between fetal brain sonography in the diagnosis of fetal ventriculomegaly (anterior horn/ 10.4mm) and dilation (combined 40.8mm). Mean Head Circumference (MHC) for neonatal hydrocephalus was 44.2cm far above the normal average of 36.7cm. When we assessed clinical benchmark of 10mm or (>19mm) for the neonate ventricle, it was far above normal range (greater than > 95th percentile value).

Key words: transfontanelle, ultrasound, CSF, brain, fetal

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Introduction

A report in Hungary revealed about 45% of the neonates with ventriculomegaly had a form of congenital infection with toxoplasmosis and cytomegalovirus accounting for roughly 70% of these cases [1]. Other studies from Europe and America reported the ventricular atrium within the 10 to 15mm range (among fetuses) or even zero incidence [2] of congenital infection. After birth there is sparse literature to support normal born neonates will have additional problems with age concerning ventricular dilation [3]. Despite numerous data on hydrocephalus, ventriculomegaly (ventricle dilation) pediatric information remains incomplete and literature are of poor quality. Some studies have researched sonar diagnosis and outcome in fetuses with Dandy Walker Malformation (DWM) [4,5].

Dandy Walker Malformation is a multiple combination of cystic dilation of the cerebellar-vermis and

larger than normal tentorial / posterior fossa [6] diagnosis proceeds from the 18th week of gestation. Ultrasound diagnosis for fetal anomaly is usually done around 18 – 23 weeks gestation. Axial diameter above 10mm across the atrium anterior /posterior horn of the lateral ventricles defines dilated ventricles (ventriculomegaly) [7]. In the past ventriculo-hemisphere ratio was used for diagnosis; grey-scale image contained skull outline lateral to the borders of posterior and anterior horns proximal to the "polygonal-shaped" cavum of septum pellucidum. Severity of Dandy Walker "cystic formation" depends on the degree of dilation and pressure of extracerebral and associated cerebral abnormalities [8,9]. Ventricular dilation is a fairly common prenatal cranial abnormality[10,11]. Atrial diameter greater than 10mm is known as ventriculomegaly [12,13]. Hydrocephalus terminology is used for ventricles with an atrial diameter greater than 15mm with rapid increase in size from onset. Other literature attempt to classify the level of ventriculomegaly [14, 15] and whether it decreases, aggravates or resolves [16] undoubtedly; normal outcome for the child may be compromised.

Case Reports

Sonographic evaluation was performed by using the Volusion E-8 (General Electric Medical Systems, Zipf-Austria) with a high frequency 7.5MHz ultrasound transducer for the neonate while a 3.5 MHz was employed for the maternal/fetal sonography. Ethical approval was granted by the Crystal Specialist Hospital and informs consent of the pregnant woman was sought and obtained in line with the 1975 Helsinki Declaration on patients' rights. The gravid woman at 23 weeks gestational age came to the radiological department for routine obstetric antenatal ultrasound while the hydrocephalic toddler was from an outpatient referral. No statistically significant asymmetry was observed between the RT and LT hemispheric halves. In the normal cephalic region, sonar level for measurement was the anterior fontanelle; as it's a constant anatomical landmark for many months in early growth percentile as described by Dewbury and Aluwihare[17].

Ultrasound was performed in neonate dorsal decubitus position while on mother's thigh; scanning time lasted for about 17 minutes. No evidence of Pascual-Castroviejo Syndrome or vermian dysgenesis seen on ultrasound. Oral interaction with the other gravid patient revealed no history of consanguineous union with her spouse. DWV was defined as complete or partial absence of the cerebellar vermis but with a small cerebellar hemisphere (though sometimes normal sized). What made this ultrasound scanning a little more "comfortable" was that it was not performed in an incubator.

The sonographic criteria for the diagnosis of ventriculomegaly (hydrocephalus) are:

- Anechoic visualization of the third and fourth ventricles with fluid
- Choroid-plexus to "dangle" with the ventricular trium
- Over-imposing ventricular boundary notices in the occipital horn and trigon areas.

Indications for neonatal cranial sonography

- Bleeding head / physical trauma
- Meningitis meningocele and convulsions
- Increased intracranial pressure
- Macrocephaly, hydrocephalus and microcephaly
- Hypoxia and distorted fontanelles

Transfontanelle ultrasound was found to be indispensable in confirmatory diagnosis of infant hydrocephalus. Mid-coronal plane measurements were taken that showed the diameter of the anterior horn of the lateral ventricles at the level of Foramen of Monro (Figure 3). Transfontanelle ultrasound is invaluable in viewing the abnormally large neonatal head circumference (HC); since the brain has no histological regenerative capacity, it is important to use a cheap, mobile, readily-available (non-invasive) sonar modality. According to the methods of Dewbury[18]; neonatal brain can be accessed at the membranous part of the temporal bone and the widely used anterior fontanelle; limited view can be obtained from posterior fontanelle. To avoid reverberation artifacts from obscuring cerebral hemisphere proximal to the transducer, a lateral section was frozen. Contrary with the assumptions of Bannister et al[19]; newborns with mild ventriculomegaly may not suffer increased intracranial pressure in-utero during prenatal life. Senat et al[20] concluded a 12mm ventricular dilation range as a variation of normal fetal anatomy (Figure 4). A further axial scan through the posterior fossa will show marked dilation of the cisterna magna at the point of communication with the fourth ventricle at the vermian defect.

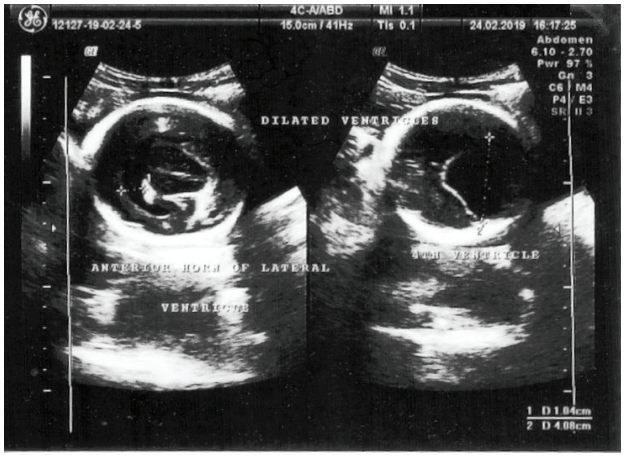

Figure 1: Sonographic measurement of fetal lateral ventricles in the axial trans-ventricular plane showing enlarged and dilated bilateral ventricles. Anatomical delineation cavum septum pellucidum was not demonstrable.

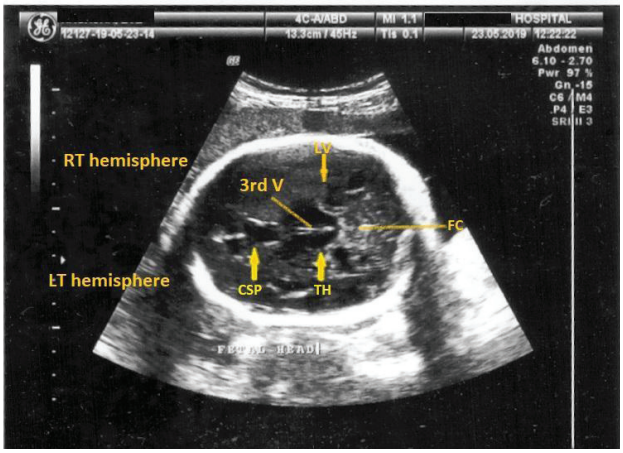

Figure 2: Transventricular ultrasound view used for measuring Head Circumference (HC) and Biparietal Diameter (BPD). Axial scan from the right occipito-anterior (ROA): a normal fetal calvaria housing the brain. Note the cavum of septum pellucidi (CSP), cerebral falx (CP), third ventricle, lateral ventricle (LV) and the thalamus (TH).

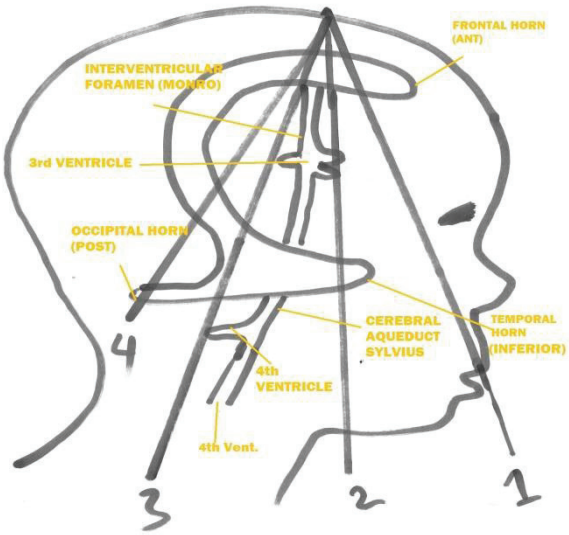

Figure 3: NOT TO SCALE- Schematic diagram representing enlarged ventricular system of a neonate in relation to transfontanelle sonar beam reverberations at various sections numbered 1,2,3 and 4.

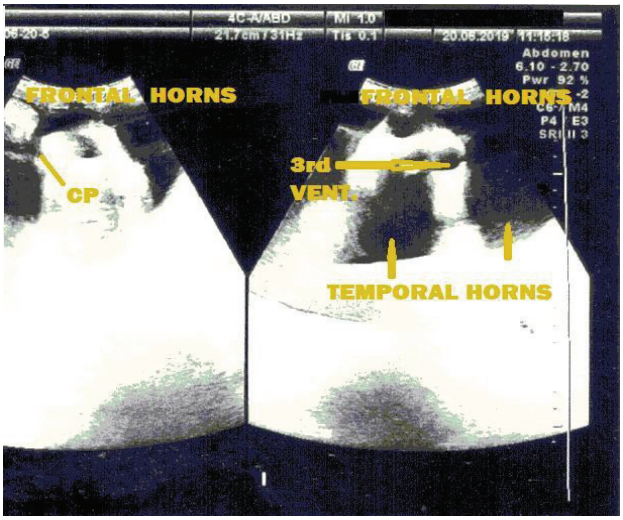

Figure 4: Ventricular Dilation: Coronal view ventricular system angled at 20° anteriorly; showing the neonatal brain and fluid, coronary plane of central 3rd ventricle was sonographically seen as a 32 mm anechoic margin. Observe the sonopenic/ echo-free superior frontal horns, inferior temporal horns and choroid plexus (CP) indentation on B-mode.

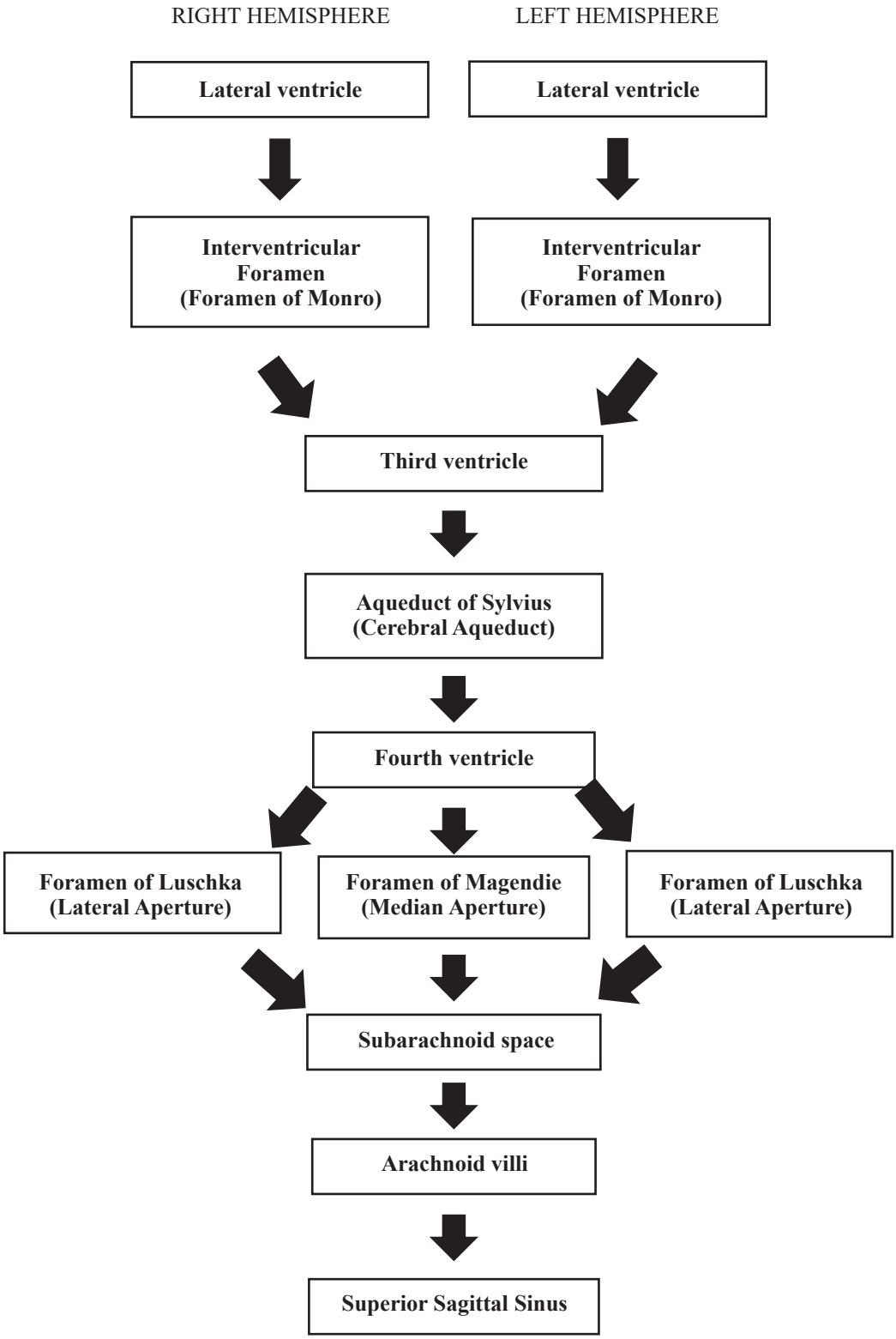

Figure 5: Flow diagram showing CSF circulation-network; obstruction in any of the part may result in any of the syndromes or anomalies described above

Discussion

The ultrasound criteria for the diagnosis of DWS in this case were particularly on complete agenesis of the vermis, cystic dilation of the 4th ventricle, upper displacement of the tentorium and an enlarged posterior fossa. Lateral ventricular diameter at the level of the Foramen of Monro was emphasized due to it being a likely region of dilatation if drainage obstruction occurs. It is important to note (to avoid a false diagnosis) that incomplete formation of the inferior cerebellar vermis and a massive 4th ventricle may give a wrong impression of vermin defect when (conclusive affirmation) is made before 16 weeks gestational age in-utero [21]. According to a research by Gilmore et al [22], there was no correlation between schizophrenia/psychosis in later years with fetal ventricular dilation. If ultrasound is not performed by a seasoned expert, it is easy to wrongly "create" the appearance of a Dandy-Walker Malformation (DWM) due to ultrasonographic reverberation errors caused by artifacts. In agreement with Gravel and Albert [23] when the ventricles are narrow, ultrasound evaluation tends to exaggerate (greater than 95th percentile) of ventricular diameter (Figure 1 and Figure 2).

It is worthy to note that any measurement used as basic threshold for the diagnosis of ventricular dilation must correlate to the plane in which the measurement is performed. Critical importance is given to the accurate determination of fetal (lateral) ventricular diameter for prenatal diagnosis of CNS diseases. Toddlers with mild fetal ventriculomegaly should be closely monitored by a pediatrician till medical professionals are confident of normal behavioral functionality. Ultrasound scan is cheap with no known adverse side effects; thus, ideal for cranial and ventricular observation. Measurements of the anterior horn could demonstrate a progressive increase up to the 6th month with marked spike in the first 90 days of post-natal life, counselling of mothers (in traditional African perspective) remains difficult; however, after parturition, neonates with associated abnormalities fare less well than babies with isolated non-progressive ventriculomegaly. Obstruction in any of the anatomical conduits with CSF (Figure 5) will result in ventriculomegaly whether it's prenatal or postnatal.

Management of the anomaly and psychological counselling of patients can be problematic and projected level of resulting handicap could be unpredictable in severe cases. In agreement with the postulation of Aletebi and Fung[24]; Dandy Walker Malformation has a greater >78% risk of developmental delay in survivors by age 4. Sagittal view via MRI may provide additional anatomical information of the vermis. In postnatal life, newborns are often observed for the evaluation of protruding fontanelle and cephalic indices greater than (>) 95th percentile (Figure 3). This is in consonance with our results; as above clinical instances, investigative diagnosis ultimately turns towards detection of ventricular dilation[25]. Female preponderance of Dandy-Walker Malformation has been described in literature[26] however contrarily; our case note is male genitalia. In toddlers who go through a transfontanelle scan, axial angle sonography may not clearly expose the cerebellar vermis. Regular ultrasound screening would not affect antenatal management and medical prognosis usually resolves with good results and viability. Medical clarity is emerging in the diagnosis and potential management of fetal ventriculomegaly; however, a problematic recurrent decimal is psychological interaction of mothers during and after pregnancy.

A recent literature by Caroll et al [27] linked a correlation between prenatal ultrasound diagnosis and pathological findings in fetal brain (anomalies). In severe Dandy-Walker Syndrome (which do not resolve), only fetuses with favorable prognosis survive. Ventriculomegaly is one of the most common ultrasound detectable fetal cerebral pathology [28]. Little data exists in literature on ventricular measurements [29,30] ; of neonatal brain at its early period [31]. Certain level of vermian dysgenesis can be found in cases with mega cistern magna; invariably having fewer posterior fossa anomalies [32].

Concerning the accuracy of fourth ventricular measurement there is a study [33] favoring access through the (coronal plane) posterior fontanelle in unresolved neonates with ventriculomegaly; considering its diameters. Diameter of the anterior horn of the RT and LT (hydrocephalic) lateral ventricles measured 15.3mm and 14.0mm respectively; far above threshold for normal (control, Figure 2) neonates RT and LT (anterior) lateral ventricles averaging 2.10mm and 2.51mm. In agreement with Thurmond et al [34] there might be associated evidence of fetal karyotyping of trisomies and decromosomal abnormalities when analyzing the genesis of ventricular dilation with (or without) mega cistern magna. Ratio of ventricular diameter to hemispheric diameter should be less than ratio 1:3; if greater as in our case (Figure 4) hydrocephalus results.

$$\frac{V.D}{H.D} \leq \frac{1}{3}$$

Conclusion

Precise measurement of the lateral ventricles is of great importance, particularly when total distance is close to 10mm threshold of fetal ventriculomegaly; contradictory classification between other radiological modalities MRI and sonography often occur. Neonates with mild ventriculomegaly should be closely monitored by a pediatrician till medical experts are confidence of normal behavioral functionality.

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