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Diagnostic

Identifying infant hydrocephalus in the emergency department with transfontanellar POCUS

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ABSTRACT

Hydrocephalus carries significant morbidity in the infant population. Although clinical symptoms are often non-specific, hydrocephalus is easily identified using transfontanellar sonography. In this review, we provide the emergency physician with a succinct overview of infant hydrocephalus and the point-of-care ultrasound (POCUS) technique for identification of this pathology.

1. Introduction

At a rate of roughly 1 in every 1000 live births, infant hydrocephalus is not uncommon, and is much higher in developing countries [1]. It is the condition most frequently treated by pediatric neurosurgeons and costs the United States 2 billion dollars in health expenditure every year. Broadly, infant pediatric hydrocephalus is categorized as congenital or acquired. Congenital causes account for the majority of infant hydrocephalus in the US, presenting at birth or developing soon after, while acquired causes are more common globally [2] (Table 1).

Initial signs of hydrocephalus can be subtle including fussiness, poor feeding, vomiting, a full fontanelle and splaying of sutures. Late findings of hydrocephalus in infants can present more dramatically with apnea, bradycardia, hypertension, lethargy, seizures, dilated scalp veins, macrocephaly and the ‘setting-sun’ sign. However, many of these listed signs and symptoms are non-specific and require simultaneous investigation of other causes such as sepsis, in tandem with an evaluation for hydrocephalus.

In the acute setting, emergency physicians often choose to obtain computed tomography of the head to evaluate for intracranial pathology, including hydrocephalus. Historically however, radiologists have used ultrasound to screen, identify and monitor hydrocephalus [1,3-5]. More importantly, if hydrocephalus is identified by ultrasound in the stable infant, an urgent MRI, rather than CT avoids radiation exposure, provides more detailed identification of cause and assists with surgical planning by neurosurgeons. For infants with ventricular shunts for hydrocephalus, an open fontanelle can provide a rapid window to evaluate the ventricles.

With the widespread use of point-of-care ultrasound (POCUS), identification of infant hydrocephalus in the ED by emergency physicians is achievable and may help with triage, flow and management decisions. In this article, we review the technique of transfontanellar POCUS, landmarks needed to recognize normal ventricles, and the sonographic features in infant hydrocephalus.

2. General scanning technique

There is no specific patient position required when performing transfontanellar POCUS. However, imaging of the brain is optimal when the infant is calm, with minimal head movement. Simple cradling by the patient's guardian, swaddling with a pacifier, warm ultrasound gel and/or use of sucrose drops as an adjunct may all help the operator keep the infant relaxed and compliant with the POCUS interrogation.

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Table 1
Congenital and acquired causes of hydrocephalus in infancy.

Congenital hydrocephalus	Acquired hydrocephalus
Anatomical malformations (primary aqueductal stenosis, foramen of Munro atresia)	Post infectious hydrocephalus (ventriculitis, meningitis)
Post hemorrhagic hydrocephalus	Neoplastic/non-neoplastic masses (ependymoma, choroid plexus cyst)
Intrauterine infections (TORCH infections)	Ventricular Shunt Malfunction
Neural tube defects (Chiari malformations, myelomeningocele)	Traumatic injury

The ventricular system of the infant brain can be visualized with transducer frequencies ranging between 5 MHz to 10 MHz. This frequency range is achievable with the phased-array, endocavitary, neonatal curvilinear, and even linear transducer [6-8]. In order to insonate the brain and visualize the ventricles, the footprint of the transducer is placed directly onto the middle of the open anterior fontanelle. The anterior fontanelle acts as an acoustic window for this POCUS application and images produced are dependent on the size of the anterior fontanelle and degree of transducer footprint contact over the open window. Although the anterior fontanelle can be open up to 2 years of age, transfontanellar POCUS is more reliable in children up to ages 12–18 months, when the anterior fontanelle is still patent, and more limited thereafter [3]. Preferably, the depth of the ultrasound beam should be adjusted to allow visualization of the base of the skull.

The infant brain is imaged in 2 planes, a coronal and sagittal view (Fig. 1). In the coronal plane, POCUS convention is to have the indicator marker point towards the right of the patient. The transducer is swept (fanned) in an anterior to posterior direction to visualize the brain in at least 3 coronal sections (anterior, middle and posterior views). In the sagittal plane, the indicator marker points to the anterior aspect of the head (face of the infant) and is swept to visualize the parasagittal left, parasagittal right, and midline of the brain.

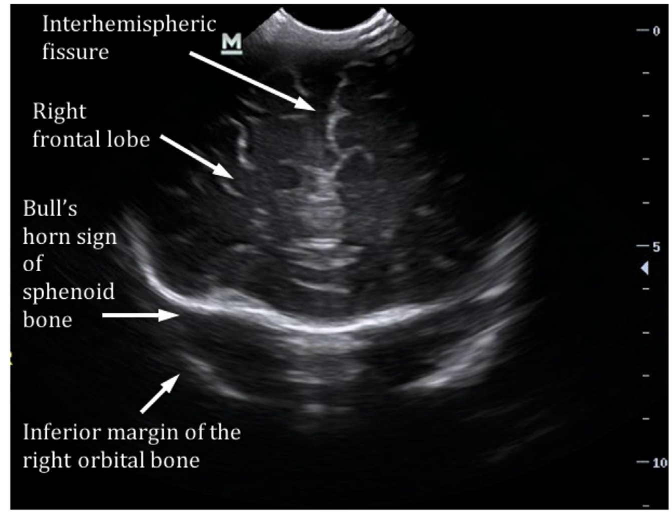

Fig. 2. Sonographic features of the anterior coronal plane.

3. POCUS image anatomy acquisition

In the coronal plane, with the indicator marker to the patient's right, the left side of the infant brain will be on the right side of the monitor. When the brain is imaged proficiently, the normal brain anatomy is virtually symmetric between left and right. Clip 1 demonstrates the images generated by sweeping the ultrasound beam anterior to posterior.

The anterior coronal section displays the frontal lobes divided by the interhemispheric fissure in the midline (Fig. 2). Other landmarks are the 'bull's horns' sign of the sphenoid bone and the superior and inferior margins of the orbits. The middle coronal section allows visualization of the body of the lateral ventricles, corpus callosum,

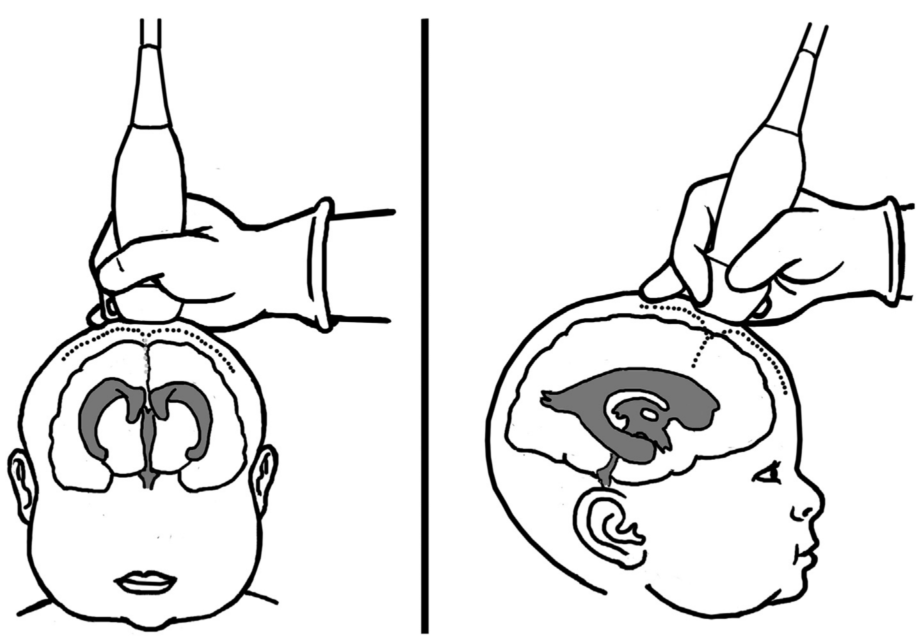

Fig. 1. Transfontanellar POCUS technique demonstrated. The left image demonstrates an ultrasound transducer with indicator marker to right of patient, allowing visualization of brain in the coronal plane. The right image has transducer with indicator marker pointing to the face of patient allowing visualization of brain in the sagittal plane.

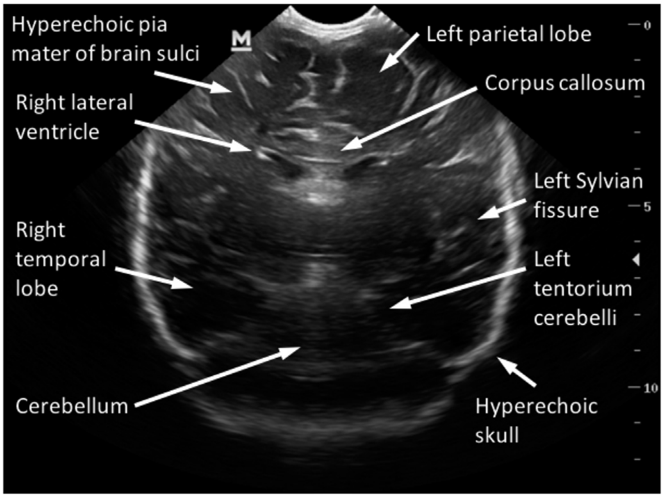

Fig. 3. Sonographic features of the middle coronal plane. The left Sylvian fissure indicated on the image separates the parietal and temporal lobe.

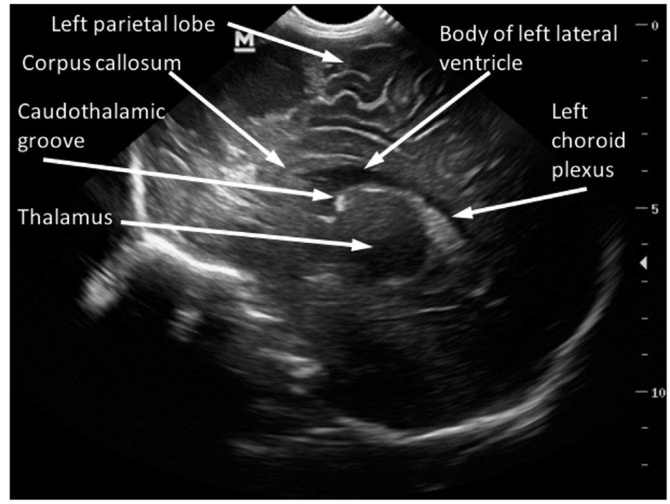

Fig. 5. Sonographic features of the parasagittal left plane.

Sylvian fissures, tentorium cerebelli, and the brainstem (Fig. 3). Typically, the temporal horns of the lateral ventricles are not visible in the coronal plane. The posterior coronal section demonstrates the region of the trigone or atrium of the lateral ventricles where the choroid plexus is most clearly visualized as a linear, oblique, hyperechoic structure bilaterally (Fig. 4).

In the sagittal plane, with the indicator marker facing the infants face, the left side of the monitor displays the anterior portion of the brain. Sweeping from left to right will bring the left ventricle, midline portion of the corpus callosum and right ventricle into view (Clip 2). The parasagittal left and right are very similar in a normal brain and usually clearly demonstrate the parietal lobe, lateral ventricle, thalamus and hyperechoic choroid plexus (Fig. 5). In infants with a wide-open fontanelle, the acoustic window allows the ultrasound wave to demonstrate the corpus callosum and third ventricle clearly in the midline sagittal plane (Fig. 6).

4. POCUS identification and interpretation of hydrocephalus

An infant with hydrocephalus is likely to have an anterior fontanelle that will allow ultrasound interrogation and visualization of the dilated ventricular system (Clips 3 and 4). There are several sonographic features that can be seen on POCUS.

An early sign of hydrocephalus is widening or 'ballooning' of the anterior horn of the lateral ventricles [9] (Fig. 7). Continued concentric dilation produces enlargement of the frontal horns with a rounded configuration, giving it a 'Mickey Mouse ears' appearance (Fig. 8). The mean and maximum diameter of the normal anterior horn of the lateral ventricle is 5 mm and 10 mm respectively. Typically, the 3rd ventricle can be visualized in the sagittal plane, although a diameter larger than 1 mm in the coronal plane is concerning for a dilated ventricular system [6]. Similarly, the temporal horn of the lateral ventricle is not visualized in a non-dilated ventricle but can be seen in hydrocephalus (Fig. 8). A temporal horn measurement of more than 3 mm qualifies the ventricle as dilated [10]. In the sagittal view the dilated body, anterior and posterior horn of the ventricle is obvious, with the choroid plexus usually appearing to dangle in the atrium of the ventricle (Fig. 9).

The bifrontal index is used in sonography as a ratio of the widest measurement of anterior horns to the widest measurement of the inner table of the skull. A ratio below 0.3 (Fig. 10) is reassuring but above 0.5 is significant and correlates to hydrocephalus [11,12] (Fig. 11). In severe hydrocephalus, significant dilation of the ventricles results in the appearance of thinning of the brain mantle.

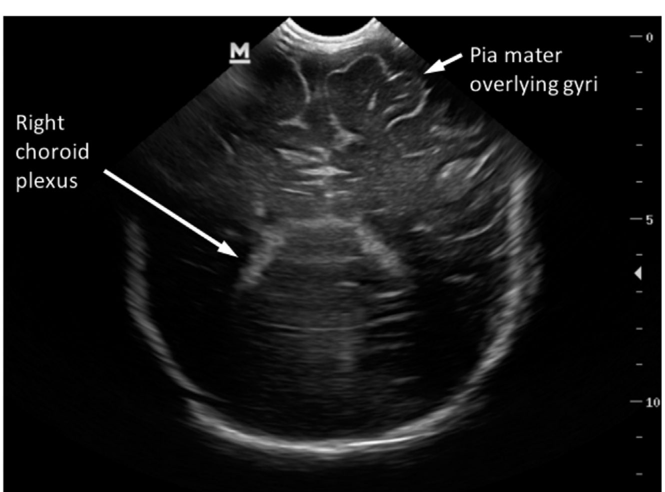

Fig. 4. Sonographic features of the posterior coronal plane.

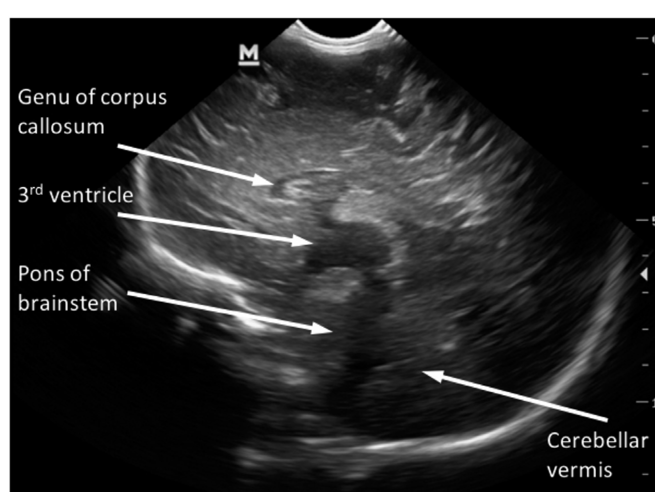

Fig. 6. Sonographic features of the midline sagittal plane.

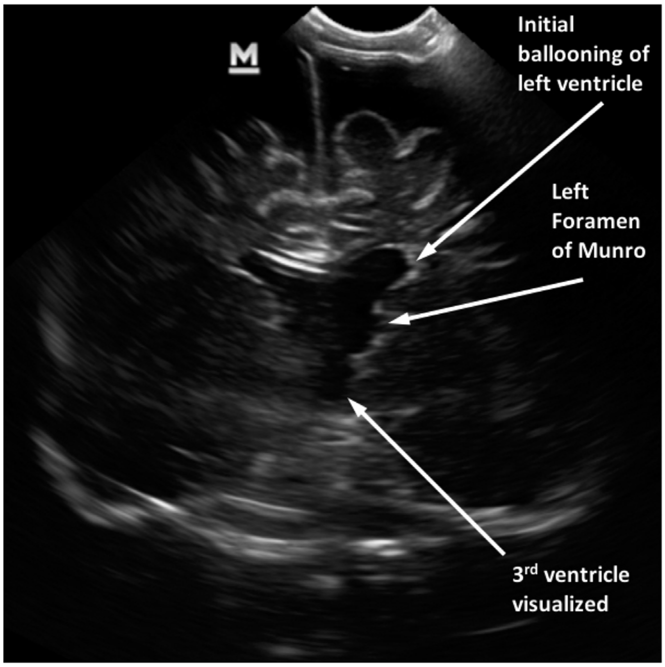

Fig. 7. Left ventriculomegaly found initially on POCUS of a 7-month-old infant with enlarging head circumference, subsequently confirmed with CT head imaging.

Overall, no one sonographic measurement is completely accurate to diagnose hydrocephalus and the constellation of sonographic findings listed above is used to help with the diagnosis.

5. Discussion

Untreated hydrocephalus results in significant morbidity. Slow increase in intracranial pressure (ICP) results in neurovascular ischemia, chronic inflammation and neuronal degeneration, impacting infant development and future cognitive function [1]. In contrast, with rapidly progressing hydrocephalus, an acute increase in ICP presents an imminent risk of intracranial herniation and mortality. Ultrasound is an established modality in the radiology department to detect this entity. Infants with an open anterior fontanelle allow the ultrasound to achieve operating characteristics approaching a sensitivity of 100% and PPV of 90% [13-16]. Ultrasound can also be used as a routine modality to monitor infants with recently placed ventriculoperitoneal (VP) shunts or when there is concern for shunt malfunction (Fig. 12) [2,17-19].

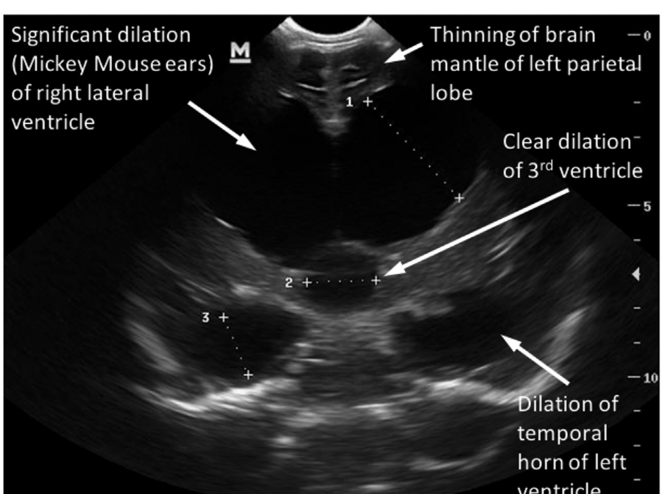

Fig. 8. 8-Month-old infant presenting with new onset hydrocephalus. Clear dilation of the ventricular system is present. 1 – Lateral ventricle measures 38 mm, 2 – 3rd ventricle measure 20 mm, 3 – Right temporal horn measures 20 mm.

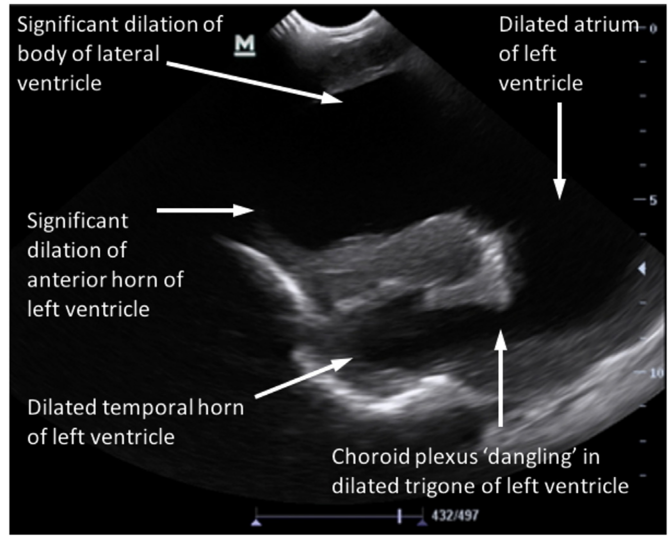

Fig. 9. – Same 8-month-old infant with dilated left ventricle seen in sagittal.

Point-of-care ultrasound has become a staple adjunct in emergency departments nationwide with both general emergency and pediatric emergency physicians utilizing it for a variety of diagnostic and procedural applications, impacting management of our patients. Pediatric emergency ultrasound fellowships have been established, and consensus guidelines have been published to provide a framework of POCUS applications in the pediatric population [20-22]. None have thus far commented on the potential scope of transfontanellar POCUS, but as pediatric emergency physicians become increasingly facile with POCUS, its application has begun to expand to include evaluation for infant hydrocephalus [7,23].

This use of transfontanellar POCUS is unique to the infant with specific complaints presenting to the ED – the vomiting child with a large head, the sleepy infant with a bulging fontanelle, the fussy infant with a VP shunt, or the neonate with a brief resolved unexplained event (BRUE). Each raises the question of hydrocephalus within the differential diagnosis, and with a wide-open fontanelle, this condition is no longer veiled to the emergency physician. The entire interrogation of the ventricular system of the infant brain can be performed rapidly, at the bedside, without need for sedation or ionizing radiation and fits well with the flow of the emergency department. Identification of hydrocephalus at the point-of-care allows quick decision making for both the community and emergency tertiary care provider alike.

6. Pitfalls

The most obvious disadvantage to transfontanellar POCUS is its reliance on the size of the anterior fontanelle. The smaller the anterior fontanelle, the smaller the window to visualize the ventricular system reliably. Furthermore, as the infant's skull size grows with age, the width of ultrasound beam is less likely to insonate the lateral and distal portions of the brain well. This is especially true for the posterior fossa, occipital horns, and third ventricle, resulting in less accurate interpretations of these regions.

Mild ventricular asymmetry is occasionally observed in neonates and is an area of uncertain consequence. It more commonly involves the left ventricle and is more pronounced at the occipital horn. Head position in this age group has some effect, but its influence diminishes with age [9]. The finding of mild asymmetry (difference of up to 5 mm) is of unclear clinical significance at this time. If the usual triangular configuration (in coronal cross section) of the lateral ventricles are

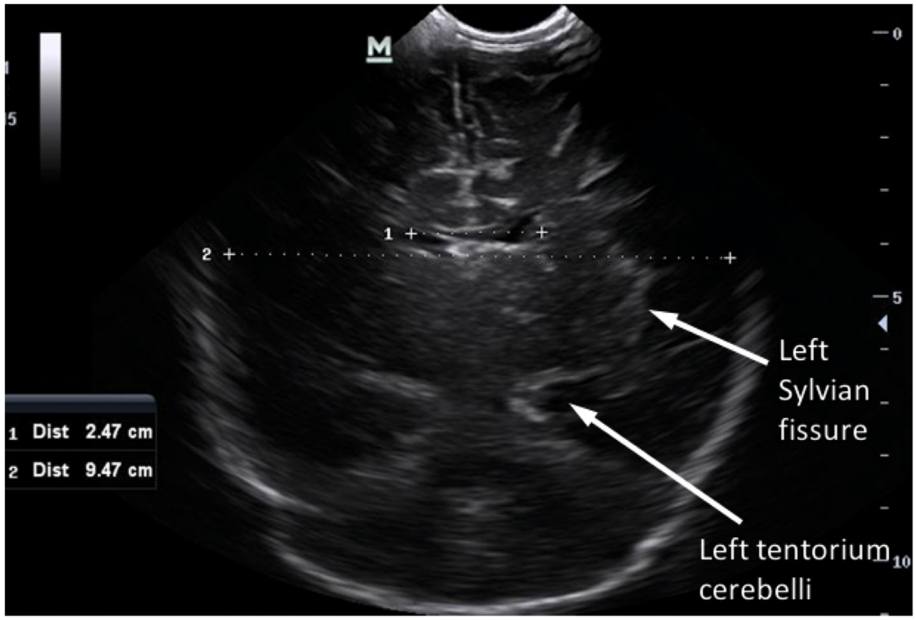

Fig. 10. Bifrontal index in a 3-month-old infant without evidence of hydrocephalus is measured at 0.2. 1 – Widest measurement of the horns of the lateral ventricles (2.47 cm), 2 – Widest measurement between the inner table of inner skull (9.47 cm).

preserved along with no other features of hydrocephalus reviewed in Section 4, non-urgent comprehensive sonography of the brain can be arranged with radiology as follow up for the patient [24].

It is important to remember that when evaluating for VP shunt malfunction with POCUS, clear images of non-dilated or baseline sized lateral ventricles (when compared to prior imaging), do not rule out shunt malfunction. Although shunt malfunction is likely if there is interval increase in ventricle size, it is still possible in normal sized or slit-like ventricles.

7. Conclusion

Transfontanellar sonography is an established method of imaging the infant brain and has long been used by radiologists to identify hydrocephalus. Emergency physicians are beginning to use this application, and are likely to find it most useful when assessing an infant with a differential diagnosis that includes hydrocephalus. In this context, transfontanellar POCUS has the ability to provide the emergency physician with a binary answer to the condition of the infant ventricular system. More research on this specific POCUS application within the field of emergency medicine is needed to determine its full potential.

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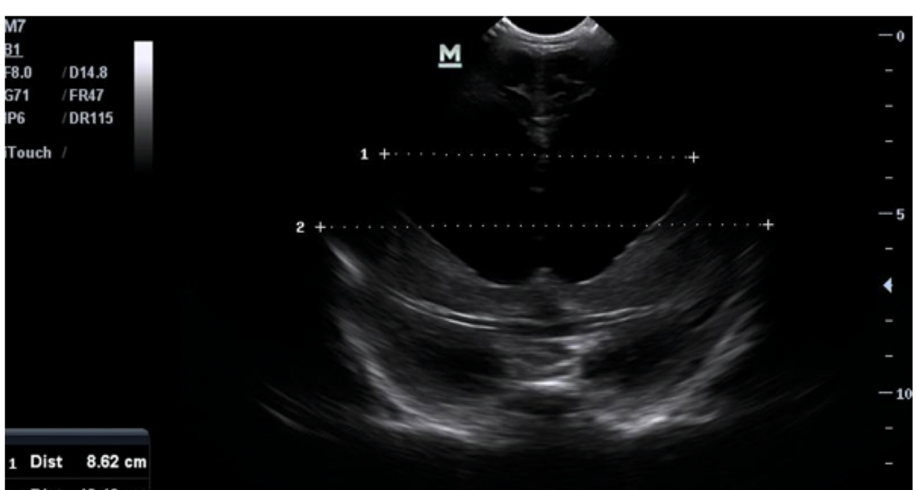

Fig. 11. Significant hydrocephalus with bifrontal ratio measured at 0.7. 1 – Widest measurement of the horns of the lateral ventricles (8.62 cm), 2 – Widest measurement between the inner table of inner skull (12.48 cm),

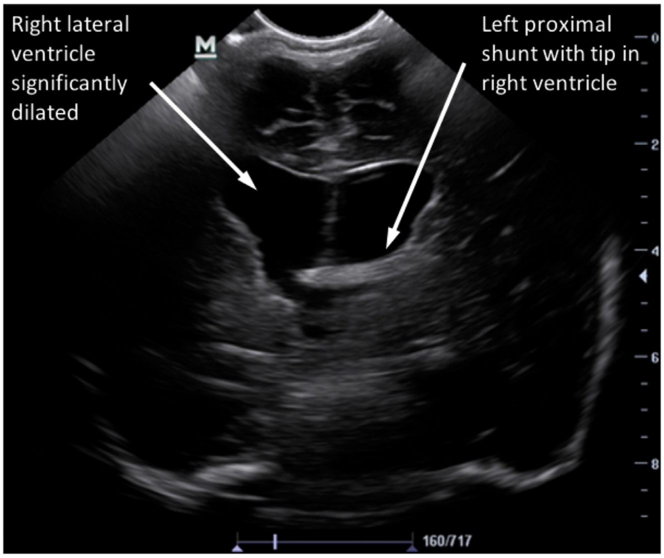

Fig. 12. Recently discharged 3-month-old presenting with fussiness 2 months after VP shunt placement. Transfontanellar POCUS revealed significant lateral ventricle dilation compared to previous CT brain. The left proximal shunt indicated in the image is seen as a hyperechoic structure with its tip ending in the right ventricle corresponding to CT brain performed after transfontanellar POCUS.

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