The "Mascara Sign": Can Hounsfield Unit Help Diagnose Proximal Ventriculoperitoneal Shunt Malfunction in Paediatric Population?

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ABSTRACT

Background: Ventriculoperitoneal (VP) shunts are prone to malfunction, often requiring multiple shunt revisions. This is associated with significant morbidity, particularly in the paediatric population. This study aims to explore whether the appearance of a proximally blocked Ventriculoperitoneal (VP) shunt can be reproducibly established, using Hounsfield Unit of attenuation as a measure of density, from non-invasive CT imaging. The benefit of such a test would be to minimise the invasiveness of shunt surgeries on patients by localising the point of fault in a malfunctioning VP shunt. **Materials and Methods:** The data set of 174 paediatric neurosurgical patients with documented proximal VP shunt blockage was identified, with 16 patients meeting the inclusion criteria. CT. imaging was reviewed with an average density obtained for each of the 16 patients. A retrospective analysis was performed using the exact Sign test to compare the median difference in average intraluminal densities using Hounsfield attenuation during proximal catheter obstruction. **Results:** There was no statistically significant median increase in intraluminal density after a proximal catheter obstruction (p=1.00). In analyzing the CT scans, we have also observed that in some patients, there is a recognisable but subjective change in appearance of the proximal catheter when blocked, the "*Mascara Sign*." **Conclusion:** Our retrospective pilot study demonstrated that Hounsfield attenuation cannot be used as an objective guide to identify proximal ventriculoperitoneal shunt obstruction on non-invasive CT imaging in the paediatric population.

Keywords: Shunt Malfunction, Shunt Revision Surgery, Hounsfield Attenuation.

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INTRODUCTION

Ventriculoperitoneal (VP) shunts are the main surgical management of hydrocephalus in both adult and paediatric populations. VP shunts are prone to malfunction, often requiring multiple shunt revision surgeries. Shunt revision rate is significantly greater in paediatric patients compared to adults and associated with increased morbidity and mortality. Most of these complications tend to occur in the early post-operative period, often within the first 6 months of shunt insertion.[1] Although multifactorial in aetiology,[2] mechanical causes account for the majority of shunt malfunctions. These include obstruction of proximal or distal catheter, valve dysfunction, shunt discontinuation (disconnection or fracture) and catheter migration. Multiple studies have identified proximal catheter obstruction as being the most common cause of shunt malfunction.[2-4]

Proximal catheter obstruction occurs due to ependymal reaction or occlusion by choroidal plexus or glial tissue, as confirmed histologically by Sekhar *et al*.[5] There is conflicting evidence as to whether choice of ventricle and attempts to optimise placement of ventricular catheter tip away from choroid plexus under radiological guidance alters the rates of proximal catheter obstruction.[6-12] Early identification of shunt malfunction in paediatric patients often poses a diagnostic dilemma to neurosurgeons. When shunt dysfunction is suspected, shunt series involving a series of plain radiographs is performed initially to exclude apparent mechanical causes. However, more detailed non-invasive imaging such as Computer Tomography (CT) scan or Magnetic Resonance Imaging (MRI) is required prior to neurosurgical interventions in order to confirm presence of hydrocephalus as well as fully evaluate the underlying mechanism of shunt failure.

Our retrospective study aims to determine whether objective radiographic variables such as Hounsfield attenuation on CT scan

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can be used as a guide to identify proximal catheter obstruction, thereby minimising the need for invasive procedures in an already challenging and vulnerable population. To our knowledge, this is the first study evaluating a potential association between Hounsfield attenuation and VP shunt patency.

At surgical removal of proximally blocked shunts, our team had repeatedly noted that there was evidence of debris, predominantly choroid plexus within and extruding from proximal shunt catheter. Upon analysis of patient scans, we have identified that in some patients, this presents characteristically on CT imaging, which we would propose as the "*Mascara sign*."

MATERIALS AND METHODS

A database of 174 paediatric neurosurgical patients who underwent shunt revision surgery from the Royal Hospital for Children and Young People (RHCYP), reviewed. Our centre serves as a tertiary neurosurgical referral centre for the South-East Scotland.

The data was categorised according to aetiology of shunt malfunction as reported on operation notes. Patients were included in the study if a proximal VP shunt obstruction was formally documented on operation notes, as well as two pre-operative CT scans, obtained before and during proximal shunt block, were available in their medical records. Delayed revision surgeries (occurring >2 days after presentation) and patients with missing operation notes were excluded from the dataset to maintain uniformity. 16 patients met the final inclusion criteria (Figure 1). To ensure patient confidentiality, data was only made accessible to individuals involved in analysis.

The CT scans were reviewed by a senior neurosurgical registrar. All CT images were analyzed using Carestream Vue PACS (Picturing Archiving and Communication System), with 3D Multiplanar Reconstruction (MPR), corrected to show lumen of catheter and peripheral in maximal continual length to remove CT bias. All images were fine cuts of at least 150 on each sequence.

Setting F3 for bone tissue, Hounsfield units were obtained at seven points each from intra-and extraluminal ventricular catheter and an average calculated to compare lucency in the ventricular catheter and wall, both prior to and during block.

Thereafter, the data set was tested for normality and the exact Sign test performed to compare the median difference in average intraluminal densities using Hounsfield attenuation during proximal catheter obstruction. For all comparisons, p-value of <0.05 was considered statistically significant. Statistical analysis was performed on Statistical Package for Social Sciences version 24 (IBM SPSS Statistics 24).

RESULTS

Aetiology of Shunt Malfunction

The aetiology of shunt malfunction (Table 1), as documented on operation notes, were proximal shunt blockage (n=55, 31%), distal blockage (n=16, 9%), valve dysfunction (n=28, 16%), shunt discontinuation (disconnection/ fracture) (n=34, 19%) and others (n=28, 16%). 22 (13%) patients were excluded due to missing or incomplete operation notes. In some cases, There was more than one underlying cause of shunt malfunction identified. The mean age of patients at time of surgery with proximal blockage was 179.66 months (range 3-261); M:F 2:1, distal blockage 162.62 months (range 120-320) M:F 2:1, valve dysfunction 110.64 months (range 11-250) M:F 2:1, shunt disconnection/fracture 153.19 months (range 22-363); M:F 2:3 and other causes 177.25 months (range 27-322); M:F 4:3.

Clinical Presentation

The most common presentation in patients with proximal shunt block (Table 2) were vomiting (n=23, 42%) and headache (n=22, 40%). Other presenting features included lethargy (n=11, 20%), drowsiness/confusion (n=9, 16%), ocular symptoms (n=8, 15%), agitation (n=5, 9%), peri-shunt swelling (n=4, 7%), bulging/full fontanelle (n=3, 5%), reduced feeding (n=3, 5%), limb weakness (n=2, 4%) and seizures (n=1, 2%). Most patients had more than one presenting feature.

Imaging Modality

For the 55 patients with proximal shunt blockage, radiological imaging obtained prior to shunt blockage were CT scan (n=38, 69%), MRI (n=11, 20%) and USS (n=4, 7%). Radiological imaging obtained during blockage were CT (n=35, 63%) and MRI (n= 12, 22%). There was no imaging available in medical records for 8 (15%) patients (Figure 2). A total of 25 patients had two pre-operative CT scans, obtained prior to and during proximal shunt block, available on Carestream Vue PACS and were thus included in the final inclusion criteria.

Hounsfield Attenuation

Average intraluminal density prior to proximal shunt obstruction was 984.00 HU (SD 223.05). Average intraluminal during proximal shunt obstruction was 986.75 HU (SD 125.668). Average extraluminal densities, i.e., tubing density measured on scans taken prior and during block were 1087.65 HU and 1117.70 HU (SD 273.865) respectively, and therefore as expected, tubing density was largely unchanged (Figure 3, Figure 4). Using the Sign test, we established that there was no statistically significant median increase in intraluminal density after a proximal catheter obstruction (p=1.00).

During CT scan analysis, it was observed that in some patients, there is a recognisable but subjective change in appearance of the

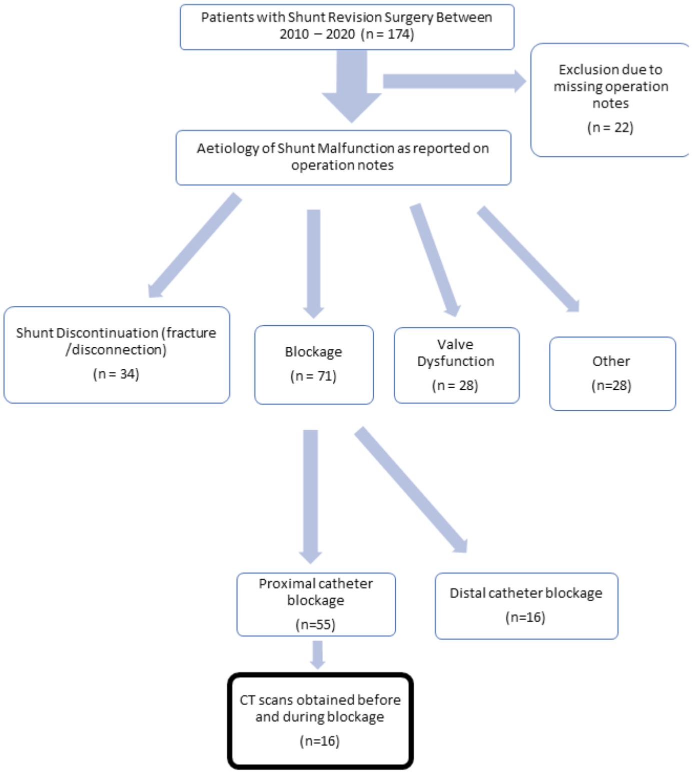

Figure 1: Diagram of selection process of data.

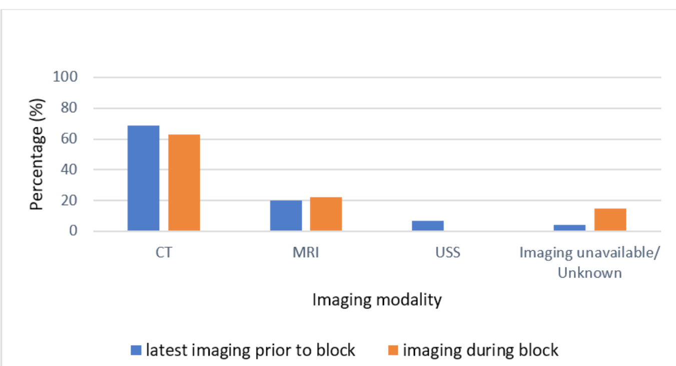

Figure 2: Proportion (%) imaging modalities used for patients (n=55) with proximal shunt blockage, prior and during blockage.

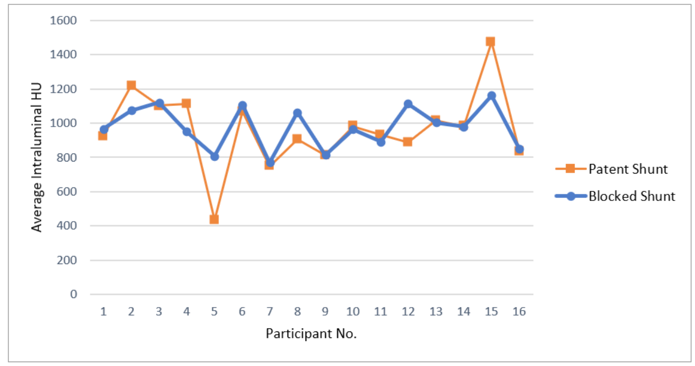

Figure 3: Line chart demonstrating the comparison between average (avg) intraluminal Hounsfield attenuation in patent versus blocked proximal VP shunt amongst the 16 participants.

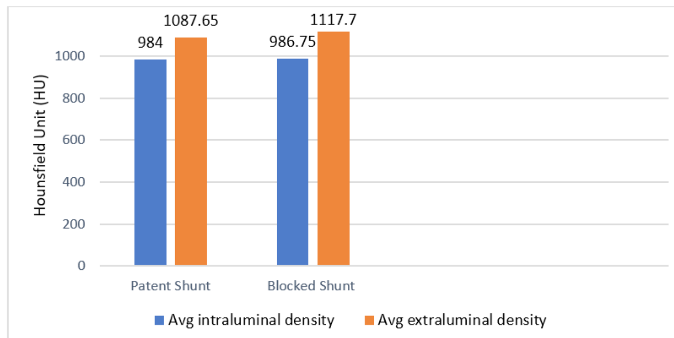

Figure 4: Bar chart demonstrating average intraluminal and extraluminal density in patent versus blocked shunt.

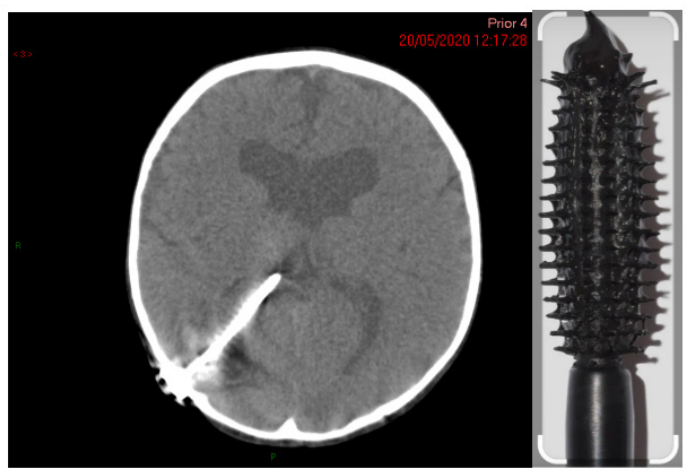

Figure 5: (Left) Mascara Sign observed on CT image analysed on Carestream Vue PACS corrected with 3D MPR to show lumen of proximal catheter; (Right) Image of mascara brush for comparison.

proximal catheter when blocked, which we would describe as the "*Mascara Sign*" (Figure 5).

DISCUSSION

The frequency of VP shunt malfunction has been reported between 45% to 59%[3,13,14-16] in various studies. The aetiology is often multifactorial.

Catheter obstruction is the commonest cause of VP shunt malfunction, as reported in previous literature.[17-21] In the paediatric population, the obstruction occurs most commonly in the proximal ventricular catheter. Our study has confirmed this. We have found 55% of all ventriculoperitoneal malfunctions were due to proximal blocks within our initial sample size of 174 participants. Similarly, to previous studies, there was also a male preponderance noted for shunt malfunction requiring revision surgeries.[22]

This study has identified that the most frequent clinical presentations of paediatric shunt malfunction reflect those reported in various other studies.[19,23,24,25] Headache (n=22, 40%), vomiting (n=23, 42%), drowsiness (n=9, 16%) and lethargy (n=11, 20%) are the common clinical predictors of shunt malfunction and may therefore indicate need for shunt revision. Atypical

Table 1: Proportion of patients and corresponding demographics of given aetiology of shunt malfunction.

Aetiology	N (%)	Mean Age (months)	Sex (M:F)
Proximal Obstruction	55 (31)	179.66	2:1
Distal Obstruction	16 (9)	162.62	2:1
Valve dysfunction	28 (16)	110.64	2:1
Discontinuation (fracture/disconnection)	22 (13)	153.19	2:3
Other	28 (16)	177.25	4:3

Table 2: Proportion of patients and corresponding mean age of given clinical presentation of proximal shunt obstruction.

Clinical Presentation	N (%)	Mean Age (months)
Vomiting	23 (42)	87.43
Headache	22 (40)	113.18
Lethargy	11 (20)	77.64
Drowsiness/ Confusion	9 (16)	89.89
Ocular symptoms	8 (15)	78.00
Agitation	5 (9)	92.00
Peri-shunt swelling	4 (7)	41.25
Bulging/ full fontanelle	3 (5)	8.33
Reduced feeding	3 (5)	107.00
Limb weakness	2 (4)	188.50
Seizure	1 (2)	84.00

presentations include seizures, ocular symptoms and a bulging fontanelle.

Shunt obstruction is diagnosed using a combination of shunt series, CT or MR imaging, and lumbar puncture. Poor Cerebrospinal Fluid (CSF) flow during a shunt tap is highly predictive of proximal shunt obstruction. Radionuclide techniques can be used to assess shunt patency as well as identify exact site of obstruction, however this is a time-consuming and expensive technique.

Modern ventriculoperitoneal shunts consist of a proximal catheter, valve and reservoir, and a distal catheter. Radionuclide techniques include injecting Technetium-99 albumin colloid or Tc-99m Diethylenetriaminepentaacetic Acid (DTPA) proximal to the reservoir and then recording activity of contrast as it passes through the shunts and reservoir. In the event of a proximal obstruction, there is reduced uptake within the ventricles.

Proximal catheter obstruction is more common than distal/ peritoneal catheter blockage. Possible causes are hypothesized as being brain parenchyma or debris such as blood and proteinaceous fluid. However, neither placement of proximal catheter within lateral ventricle farthest from the choroid plexus[18] nor analysis of CSF samples to exclude hemorrhage and infection[25-27] have shown any association with shunt survival. Nevertheless, literature reports that "good" interventricular positions for the catheter tip are the third ventricle, foramen of Monro and lateral ventricle and are associated with the best shunt survival. On the contrary, "bad" positions are walls of the lateral ventricle and septum pellucidum and are most often associated with postoperative shunt revision.[28]

CT brain has been the most widely used investigation and is considered the primary investigation by most.[2,19,29] Over the years, many studies have attempted to use various radiological indices or parameters of non-invasive imaging, both CT and Non-CT to identify shunt malfunction especially obstruction preoperatively but also objectively and non-invasively. Therefore, we have resorted to this widely used imaging modality to explore whether there is something we may be missing and can something as simple as Hounsfield attenuation be used to exclude catheter obstruction in an obstructive manner?

Pre-operative ventricular size on CT scan is an unreliable predictor of shunt malfunction.[30-32] Ventricles may even be dilated as patient improves clinically and contrarily, there may be slit-like ventricles in a patient with hydrocephalus secondary to shunt malfunction.[33] A rapid reduction in ventricular volume has been reported to be an indicator for requiring early shunt revision. Moreover, dilated ventricles are more often associated with distal shunt failures than proximal. Reduction in ventricular pressure better correlates to shunt efficacy than ventricular volume.[34]

This study aimed for an objective assessment for diagnosing shunt malfunction using Hounsfield attenuation as a measure of tubing density. To the best of our knowledge, there has been no prior literature evaluating an association between Hounsfield attenuation on CT scan and shunt patency. The closest study we have obtained was that conducted in Hanyang University Medical Center in Seoul, Korea evaluating a possible association between Hounsfield (HU) attenuation on CT imaging and the possible association to shunt-dependent hydrocephalus.[35] This study conjectured that lower skull BMD may correlate to poor arachnoid trabeculae integrity as both contain type I collagen. Thus, it hypothesized that low skull HU values may correlate to shunt-dependent hydrocephalus in patients that received cranioplasty after decompressive craniectomy for traumatic acute subdural haematoma. The study finds that the optimal cut-off values for mean frontal skull HU and HU at internal occipital protuberance for predicting shunt-dependent hydrocephalus were 797.375 (sensitivity = 81.8%; specificity = 586.500 (sensitivity = 78.8%; specificity = 58.1%). This lower Bone Mineral Density (BMD) group had an 8.6-fold increased risk of requiring shunt-dependent hydrocephalus. However, we acknowledge that this cannot be used for exact comparison with this study hypothesis as the mechanism of hydrocephalus secondary to shunt obstruction differs from post-hemorrhage hydrocephalus.

Limitations

This study has several limitations. The sample size was too small to draw reliable conclusions from the dataset; however, the study is reproducible on a larger sample size taking into account all the limiting factors. Despite that the Hounsfield measurements were intended to represent an objective measurement of density; this does not account for inter-scanner differences. Birnbaum BA *et al*. reported that there are variations in Hounsfield values between scanners including Philips and Siemens, however this is very minute (range 0-20 HU).[36] In addition, there is the caveat of interobserver bias whilst selecting the exact points for Hounsfield measurement. To account for this bias, In Suk Bae *et al*. recommends an oval function called 'Region of Interest (ROI)' whereby the maximum, minimum and mean HU values can be automatically obtained by the PACS system.[35] This function unfortunately, was unavailable on our electronic system. Moreover, there may be selection bias as we have excluded patients undergoing emergency shunt revision. We limited the patient population in the study to increase uniformity of the data.

CONCLUSION

To conclude, this study reiterates previous literature in that catheter obstruction is the commonest cause of ventriculoperitoneal shunt malfunction in the paediatric population. This presents as headache, vomiting, confusion and lethargy. The most common site of obstruction is the proximal ventricular catheter. CT imaging is the most common and primary investigation for suspected shunt malfunction. We have found that the average Hounsfield attenuation and therefore intraluminal density of the proximal ventricular catheter remains unchanged post-obstruction. The average extraluminal HU, which measures the tubing density, is greater than that within the catheter lumen. CT scan analysis of paediatric shunt obstructions revealed a characteristic but subjective appearance, which we would describe as the *Mascara sign*.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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