

# Pathophysiology of Isolated Lateral Ventriculomegaly in Shunted Myelodysplastic Children

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**Abstract.** Eight myelodysplastic children developed isolated lateral ventriculomegaly following shunt insertion for progressive hydrocephalus after closure of a myelomeningocele. In all patients a low-pressure distal slit valve (Uni-shunt) system preceded development of an isolated contralateral ventricle. Six of 8 children required a second contralateral shunt for a symptomatic isolated ventricle. Magnetic resonance imaging demonstrated a collapsed ventricle ipsilateral to the shunt secondary to distortion of the foramen of Monro. This was clearly depicted using three-dimensional color reconstructions of the ventricular anatomy. Low-pressure distal slit valves should be avoided in myelodysplastic children to prevent postshunt ventricle isolation.

## Introduction

Myelodysplastic children are well known to have multiple anomalies of the central nervous system and often require cerebrospinal fluid (CSF) diversion procedures following closure of the myelomeningocele defect. A series of children with this birth defect treated at our institution developed isolated lateral ventriculomegaly following placement of a ventriculoperitoneal shunt, prompting further investigation into the etiology of this finding. Although this phenomenon has been described before [1-4], the pathophysiology remains speculative. Known causes of isolated ventricles include atresia of the foramen of Monro [5-9], infections [10-12], tumors [13], intraventricular hemorrhage [14], and following ventricular shunts for nonmyelodysplastic hydrocephalus, although this rarely isolates the shunted ventricle [15].

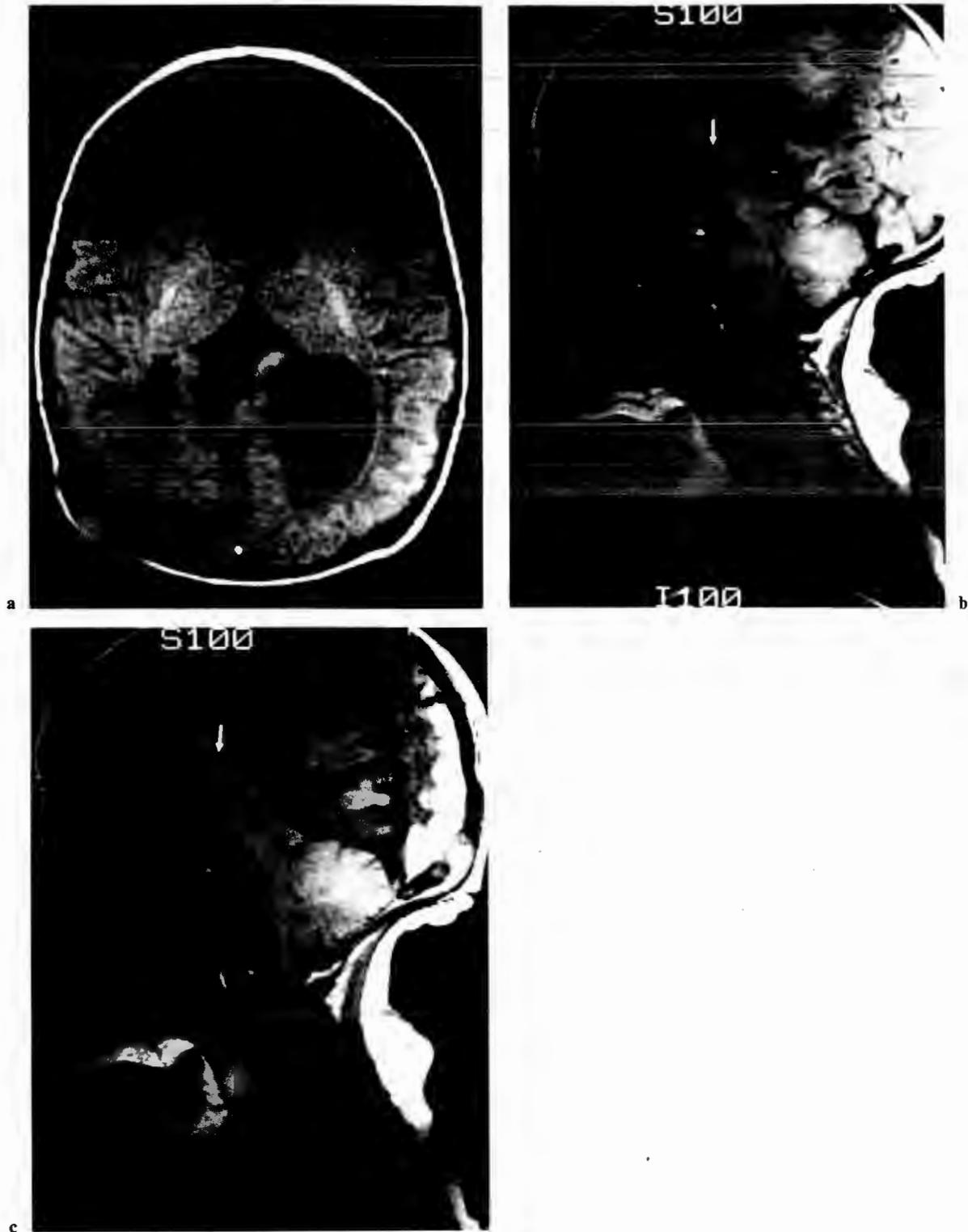
With the advent of magnetic resonance imaging (MRI) using a high field strength magnet, the ventricular anatomy may now be more clearly defined following shunt placement. In addition, computer technology exists for three-dimensional volumetric analysis of the ventricular

system which will help to further elucidate the pathophysiology of isolated ventricles. We reviewed the MRI scans of myelodysplastic children identified to have postshunt isolated lateral ventriculomegaly, and reconstructed a three-dimensional color contour of the foramen of Monro region in 1 patient to further define the mechanism and anatomy of the obstruction.

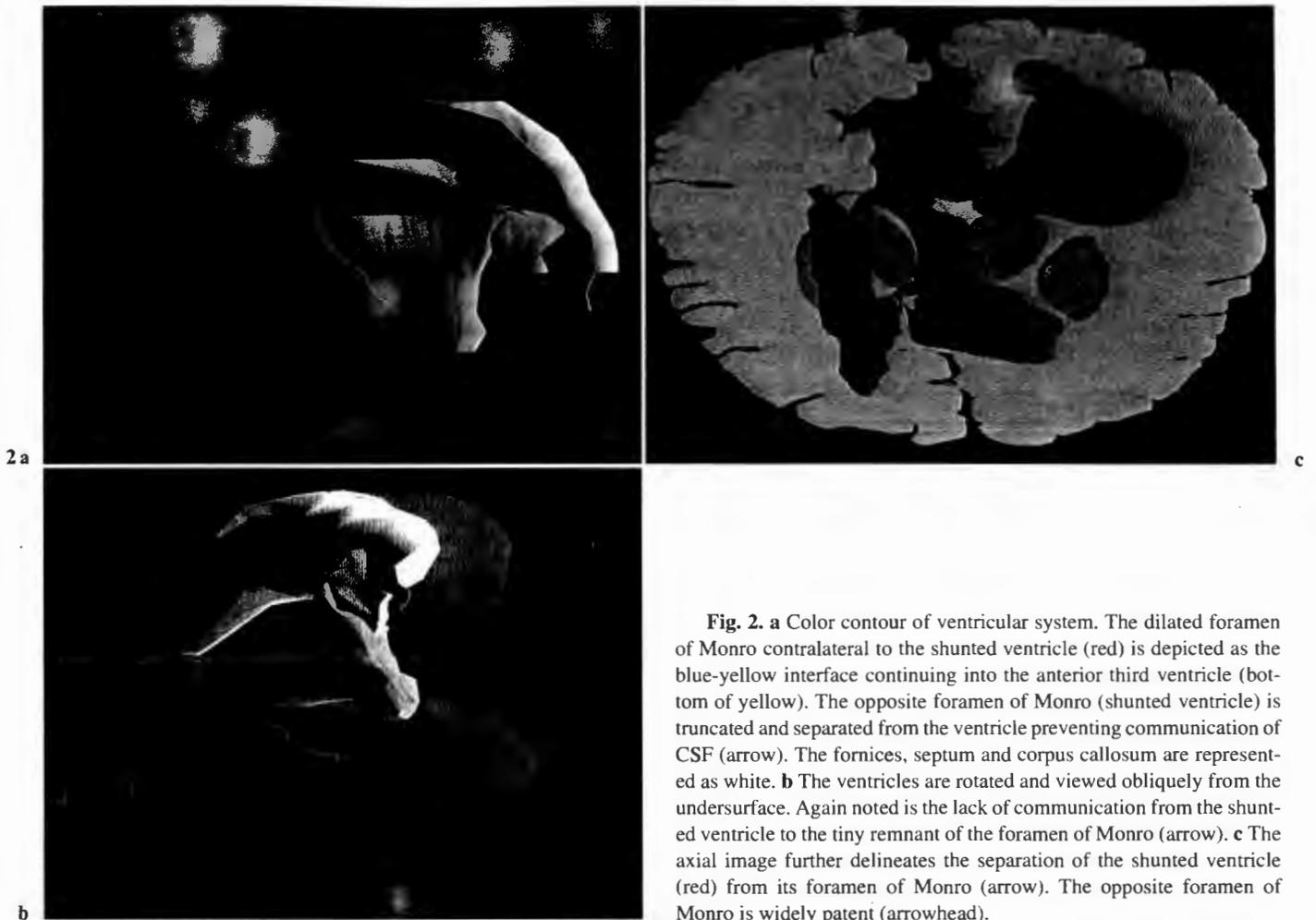
## Materials and Methods

From 1986 to 1988 inclusive, children with myelodysplasia undergoing placement of a ventriculoperitoneal shunt for hydrocephalus were retrospectively reviewed to determine the incidence of isolated ventriculomegaly following shunt placement. Eight of 44 children developed this condition whereas no patients without myelodysplasia (e.g., congenital hydrocephalus) and excluding neoplasia and infections developed an isolated lateral ventricle during the same time period. Children in this latter group with slight ventricular asymmetry following shunt placement were not classified as having isolated ventriculomegaly.

All 8 patients underwent closure of the myelomeningocele within 72 h of birth. Preoperative cranial ultrasound and computed tomography (CT) scans confirmed symmetrically enlarged ventricles. MRI



**Fig. 1. a** T-1-weighted axial MR scan at the level of the foramen of Monro. The anterior tubercle of the thalamus is large and eccentric (arrowhead) and the massa intermedia thickened (open arrow). The fornices are deviated toward the collapsed ventricle and the foramen of Monro is torqued (arrow). **b** T-1-weighted sagittal MR scan demonstrating patent foramen of Monro on the side of the nonshunted ventricle (arrow). Note the thickened massa intermedia (m). **c** T-1-weighted sagittal MR scan demonstrating obstructed foramen of Monro ipsilateral to the collapsed ventricle (arrow).



**Fig. 2.** **a** Color contour of ventricular system. The dilated foramen of Monro contralateral to the shunted ventricle (red) is depicted as the blue-yellow interface continuing into the anterior third ventricle (bottom of yellow). The opposite foramen of Monro (shunted ventricle) is truncated and separated from the ventricle preventing communication of CSF (arrow). The fornices, septum and corpus callosum are represented as white. **b** The ventricles are rotated and viewed obliquely from the undersurface. Again noted is the lack of communication from the shunted ventricle to the tiny remnant of the foramen of Monro (arrow). **c** The axial image further delineates the separation of the shunted ventricle (red) from its foramen of Monro (arrow). The opposite foramen of Monro is widely patent (arrowhead).

scans were performed in children with isolated ventriculomegaly to define the anatomical features of the region adjacent to the foramen of Monro. In addition, a three-dimensional reconstructed color image of the ventricular system was carried out in 1 case to attempt to define the pathophysiology of the isolated ventricle. The contour of the ventricular system demonstrated on thin-section (3 mm), axial T-1-weighted (TR 600, TE 20) images was manually traced onto paper sheets which were placed sequentially onto a digitizing tablet. University of Washington software has been developed to digitize contours, to combine the tables of X-, Y-, Z-coordinates into a coherent numerical description of the original object and to transform these coordinates into a format suitable for interactive computer graphics [16]. Our programs are used to produce three-dimensional anatomical reconstruction [17]. Various structures may be colored with a range of hues, and other surfaces made translucent to further delineate the ventricular system. Rotation and translation is possible to depict certain anatomical features and may be enhanced with variable lighting effects. The data is recorded on videotape or videodisc, and slides or photos are made directly from the high resolution monitor or through a camera attachment.

#### *Summary of Cases*

The first shunt was placed within 21 days following closure of the myelomeningocele (mean, 9 days). In all instances a low pressure distal slit valve (Unishunt) system was placed prior to the development of an isolated ventricle. Seven of eight shunts were placed posteriorly with the remaining shunt system placed anteriorly at the level of the coronal suture. Initially, 2 of 8 patients had a medium pressure (non-Unishunt) proximal valve shunt system placed, however, due to persistent, symmetric ventriculomegaly, underwent revision to a low pressure Unishunt.

Symptoms indicative of shunt malfunction, e.g., head circumference change, full fontanelle, irritability, etc., necessitated evaluation with a CT scan in 6 of 8 patients. However, instead of finding a malfunctioning shunt, the CT, and, subsequently, the MRI scan demonstrated the consistent finding of a collapsed ventricle ipsilateral to the shunt with a deviated and distorted foramen of Monro on the same side. The opposite ventricle was enlarged, in addition to the third ventricle, and a patent contralateral foramen of Monro was apparent (see fig. 1). All 6 of the children required a second contralateral shunt (4 low pressure, 2 medium pressure systems) due to symptomatic isolated ventriculomegaly which resulted in resolution of their symptoms. The remaining 2 patients had no symptoms referable to the isolated ventricle and were managed conservatively. Myelomeningocele patients with medium pressure shunts (proximal and distal valves) and nonmyelodysplastic hydrocephalic children with low or medium pressure shunts (proximal and distal valves) did not develop an isolated contralateral ventricle during the same period of time. However, 1 child in this latter group, e.g., nonmyelodysplastic congenital hydrocephalus, had diagnostic imaging evidence of a partially isolated ventricle that was asymptomatic. In the original group of 44 myelodysplastic infants, no child had placement of a high pressure, distal valve Unishunt.

A three-dimensional animated color contour drawing of the ventricular system was performed in 1 case (see fig. 2). By rotating the image to visualize the reconstructed collapsed ventricle it became apparent that the foramen of Monro ipsilateral to the shunt was 'torqued' in such a manner that the foraminal opening became closed. This prevented the flow of CSF from the contralateral ventricle, through its own patent foramen of Monro, into the shunted ventricle, and resulted in isolation of the contralateral and third ventricle.

## **Discussion**

Anomalies of the lateral ventricular system in myelodysplastic children have been well described in the literature. The massa intermedia, which is absent in only 17% of the general population [12], is typically enlarged in children with a myelomeningocele and Chiari II malformation [18-20]. This finding, in addition to an eccentric bulge from the head of the caudate nucleus [21], prominent commissural fibers extending across the anterior third ventricle [22], and anterior pointing of the frontal horns [22, 23], results in an abnormally distorted and narrowed foramen of Monro [24].

Following shunt placement in children with a myelomeningocele, it has been noted that unilateral dilation of the contralateral, nonshunted ventricle may occur [2, 3, 25]. Hubballah and Hoffman [26], Babcock and Han [1] and Oi and Matsumoto [25] have postulated that a functional obstruction takes place following ventricular drainage at the level of the ipsilateral foramen of Monro. An additional factor contributing to the isolated ventricles, namely the use of low pressure shunts, was also mentioned [14, 25, 26]. After reviewing the MRI scans on our patients we noted that the dimensions of the foramen of Monro were greatly distorted giving the appearance of 'foraminal torsion', yet this was difficult to visualize in static, multiplanar images. Therefore, the ventricular volume was reconstructed as an animated three-dimensional cast and rotated in multiple planes to visualize each foramen of Monro separately. By shading each ventricle with a different color scheme, we were able to demonstrate the foramen of Monro ipsilateral to the shunt demonstrating that it was shifted and contorted resulting in a complete occlusion. Although the contralateral foramen of Monro was patent, its ventricle became isolated from the shunted side due to that occlusion.

Rekate et al. [27] has postulated, based upon a dog model used to study the resistance level at the foramen of Monro, that the foramen acts as a valve creating resistance to CSF flow. Certainly, the anatomical anomalies seen in this region associated with myelodysplasia will act to increase the functional resistance and accentuate the anatomical block. Therefore, low pressure shunts, especially those systems with distal slit valves that tend to overdrain the ventricles, should be avoided in newborn babies with myelodysplasia due to the possibility of torsion and occlusion of the foramen of Monro ipsilateral to the shunt. Not all patients with myelodysplasia and low pressure shunts will develop this condition due, most likely, to the often recognized finding of an incompetent septum pellucidum

[4, 22]. Therefore we currently recommend first revising the shunt to a medium pressure (proximal or distal valve) system or adding an antisiphon device if the child is symptomatic with an enlarged contralateral ventricle, versus placement of a (second) shunt into the isolated ventricle. Avoidance of a low pressure shunt with a distal slit valve should prevent isolation of the lateral ventricle in shunted myelodysplastic children.

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