



Craniomegaly in Neonate and Infants Requiring Neurosurgical Intervention: An Experience at Tertiary Care Center

Sangh Mittra¹ Hanuman Prasad Prajapati² Raj Kumar²

¹Department of Gynaecology, Veerangana Avanti Bai Mahila Chikitsalaya, Lucknow, Uttar Pradesh, India

²Department of Neurosurgery, Uttar Pradesh University of Medical Sciences (UPUMS), Saifai, Etawah, Uttar Pradesh, India

Address for correspondence Raj Kumar, MCh, Department of Neurosurgery, Uttar Pradesh University of Medical Sciences (UPUMS), Saifai, Etawah 206130, Uttar Pradesh, India (e-mail: rajkumar1959@gmail.com).

Indian J Neurosurg 2023;12:42–46.

Abstract

Background The identification of neurosurgical causes of craniomegaly and early institution of therapy requires for better clinical and functional outcomes.

Aims and Objectives The aim of this study was to evaluate the neurosurgical causes, managements, and outcomes of craniomegaly in neonate and infants.

Materials and Methods The cases with a history of head enlargement from neonatal period were included in this study. Their causes, managements, and outcomes were recorded retrospectively during the period of January 2010 to February 2013, in neurosurgery department at SGPGIMS Lucknow, and June 2018 to June 2020, at UPUMS, Saifai, Etawah, UP, India.

Results Out of 41 cases, there were 30 (73.14%) cases of hydrocephalus, 4 (9.76%) Dandy-Walker malformation, 2 (4.88%) subdural collection, 2 (4.88%) arachnoid cyst, 1 (2.44%) craniosynostosis, and 2 (4.88%) with tubercular meningitis. The age range of our cases was 18 to 178 days and the mean age was 102.54 ± 50.73 . Preoperative head circumference range was 39 to 62 cm (mean: 55.27 ± 6.58 cm). Majority of the cases ($n = 32$, 78.05%) were managed with ventriculoperitoneal shunt surgeries. Out of 41 cases, 33(80.49%) had improved outcomes, 7 (17.07%) stabilized, and mortality occurred in 1 (2.44%) case. Postoperatively, there was improvement in the head circumference (range: 39–60 cm and mean: 46.15 ± 5.83 cm) on 6 to 24 months (mean: 17.85 ± 5.18 months) of follow-up.

Conclusion Hydrocephalus was the commonest neurosurgical cause of head enlargement in neonate and infants. Shunt surgery was the most common form of management of these cases. Early detection, institution of therapy, and periodic follow-up program for diagnosing and treating complications were the key to successful outcomes in these patients.

Keywords

- craniomegaly
- newborn and infants
- etiologies
- management and outcomes

article published online
December 17, 2021

DOI <https://doi.org/10.1055/s-0041-1735377>.
ISSN 2277-954X.

© 2021. Neurological Surgeons' Society of India. All rights reserved. This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)
Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

Introduction

Craniomegaly (head enlargement) is also known as macrocephaly. It is defined as head circumference above the 98th percentile or greater than 2 standard deviation above the mean for the given age, sex, race, or gestational age.¹ The underlying structures that may lead to craniomegaly include the scalp, cranial bone, and the contents of the cranium. Benign familial macrocephaly, an autosomal dominant trait, is the commonest cause of craniomegaly.² Craniomegaly can be sorted into genetic or nongenetic, congenital or acquired, pathological or idiopathic, syndromic or nonsyndromic (isolated), and several other varieties.

Incidence of isolated craniomegaly is found to be 0.5%.³ The incidence increases in populations with mental retardation, autism, or associated malformations. It may be associated with a constellation of other anomalies as part of recognized syndromes.⁴ Studies regarding the neurodevelopmental and neuropsychological profile of patients with craniomegaly show contradictory results, ranging from normal cognitive and motor functioning to substantial neuropsychological disabilities.⁵ It is also not clear whether there is a difference in prognosis between patients with isolated craniomegaly and those associated with other brain or systemic anomalies.

On the basis of physical, metabolic or brain imaging findings craniomegaly can be categorized into various subtypes.⁴ Out of these, few may need urgent attention and management to avoid the harmful effect on growing brain. These includes hydrocephalus (congenital/ acquired), subdural collections (SDCs), craniosynostosis, Dandy-Walker malformation, and arachnoid cysts.

We conducted a retrospective study to investigate various aspects of this common neurosurgical condition, their etiology, presenting complaints, clinical findings, treatments, and outcomes.

Materials and Methods

It was a retrospective study done during the period of January 2010 to February 2013 at neurosurgery department of Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, and June 2018 to June 2020, at Uttar Pradesh University of Medical Sciences (UPUMS) Saifai, Etawah, UP,

India. All the cases with a history of head enlargement from neonatal period were included in this study irrespective of their age of presentation.

Inclusion Criteria

Only the patients needing neurosurgical care were included in this study such as:

1. Intracranial expansile conditions like hydrocephalus—congenital/acquired, SDCs, Dandy-Walker malformation, and arachnoid cysts.
2. Craniosynostosis, that is, scaphocephaly.

Exclusion Criteria

The following cases were not included in our study

1. Benign familial macrocephaly
2. Autism-associated macrocephaly
3. Genetic causes of macrocephaly
4. Metabolic causes of macrocephaly
5. Pediatric brain tumors
6. Cases with inadequate records
7. Cases with follow-up less than 6 months

All the cases were thoroughly investigated and offered defined standard treatments as per departmental protocol. Details of the treatments given are shown in ►Table 1.

Ommaya procedure was done in 1 (2.44%) case with the diagnosis of hydrocephalus (aqueductal stenosis) in view of patient poor clinical condition. Later on ventriculoperitoneal shunt (VPS) had been put in the same case. In the SDC group, both the cases were kept on conservative line of treatment initially, but burr hole drainage required in one case later. The cases with arachnoid cyst and craniosynostosis (scaphocephaly) were managed conservatively.

Shunt revision surgery was required in six cases (5 VPS and 1 cystoventriculoperitoneal shunt [CVPS]). Out of five VPS revision cases, there were four cases of aqueductal stenosis and one case of tubercular etiology group. In all the five cases, the cause of shunt failure was blockage. One VPS case was revised 2 years after the primary procedure, rest four VPS were revised within 1 month after the first operation. In Dandy-Walker malformation cases, one had cerebrospinal fluid (CSF) leak after CVPS. We had revised CVPS twice due to blockage at proximal end.

Table 1 Details of surgical treatments

S. no.	Treatment	HCP (n = 30)	DWM (n = 4)	SDC (n = 2)	AC n (%)	CS n (%)	TBM n (%)
1	VPS	28 (68.29%)	1 (2.44%)	0	0	0	2 (4.88%)
2	Ommaya procedure	1 (2.44%)	0	0	0	0	0
3	CVPS	0	2 (4.88%)	0	0	0	0
4	Burr hole	0	0	1 (2.44%)	0	0	0
5	Conservative	1 (2.44%)	1 (2.44%)	2 (4.88%)	2 (4.88%)	1 (2.44%)	0
6	Shunt revision	4 (9.76%)	1 (2.44%) (CVPS)	0	0	0	1 (2.44%)

Abbreviations: AC, arachnoid cyst; CS, craniosynostosis; CVPS, cystoventriculoperitoneal shunt surgery; DWM, Dandy-Walker malformation; SDC, subdural collection; TBM, tubercular meningitis; VPS, ventriculoperitoneal shunt surgery.

Table 2 Distribution of etiology according to age and sex

Age groups (in days)	HCP n (%)	DWM n (%)	SDC n (%)	AC n (%)	CS n (%)	TBM n (%)	Total n (%)
0–30	4 (9.76%)	1 (2.44%)	0	0	0	0	5 (12.20%)
31–60	4 (9.76%)	0	0	0	1 (2.44%)	0	5 (12.20%)
61–90	5 (12.20%)	0	0	1 (2.44%)	0	0	6 (14.63%)
91–120	8 (19.51%)	0	1 (2.44%)	0	0	0	9 (21.95%)
121–150	2 (4.88%)	1 (2.44%)	0	0	0	2 (4.88%)	5 (12.20%)
151–180	7 (17.07%)	2 (4.88%)	1 (2.44%)	1 (2.44%)	0	0	11 (26.83%)
Male	24 (58.54%)	2 (4.88%)	1 (2.44%)	1 (2.44%)	1 (2.44%)	1 (2.44%)	30 (73.17%)
Female	6 (14.63%)	2 (4.88%)	1 (2.44%)	1 (2.44%)	0	1 (2.44%)	11 (26.83%)
Total	30 (73.17%)	4 (9.76%)	2 (4.88%)	2 (4.88%)	1 (2.44%)	2 (4.88%)	41 (100%)

Abbreviations: AC, arachnoid cyst; CS, craniosynostosis; DWM, Dandy-Walker malformation; HCP, hydrocephalus; SDC, subdural effusion; TBM, tubercular meningitis.

In aqueductal stenosis group, one case had vomiting 1.5 month after surgery and other had seizure 1 year after the ventricular shunting. Both of these cases responded to conservative treatments. One case of postinfectious hydrocephalus group had ptosis and diminution of vision at 2 and 10 months, respectively, at follow-up following surgery.

On each follow-up, serial neurological examination, developmental evaluation, head-circumference measurement, and radiological assessment were done in all cases. Initially, all the children were followed up at 6 weeks, then at an average of 3 months, 6 months, and at yearly interval until they had some complication. Outcomes of craniomegaly were measured clinically and functionally as improved (reduction in head circumference and improvement in the clinical status after surgery) and stable (head circumference remains same as in preoperative states) at 6 to 24 months (mean: 17.85 ± 5.18 months) of follow-up.

Statistical Analysis

Data work entered in Microsoft Office Excel 2007 and analyzed using SPSS version 24.0 (IBM Corp.; Chicago, Illinois, United states). Data were analyzed at two levels: descriptive and analytical. Frequency, percentage, range, means, and median were used to describe the characteristics of study participants.

Results

Over the given period, total 66 cases with head enlargement were admitted and investigated. Out of 66 cases, 41 fulfilled the inclusion criteria's and selected for this study. All the cases experienced symptom onset during the first month of life, whereas age of presentation to the hospital varied. The age range of our study cases was 18 to 178 days (mean \pm standard deviation = 102.54 ± 50.73 days). There were 30 (73.17%) males and 11 (26.83%) females. Majority ($n=30$; 73.17%) of the cases of craniomegaly were secondary to hydrocephalus. Most of the cases ($n=11$, 26.83%) were in

age group of 151 to 180 days. Age and sex wise distributions of etiologies were given in **Table 2**.

Increased head circumference was the common clinical feature in all the 41 cases. The preoperative head circumference range of these children was 39 to 62 cm (mean: 55.27 ± 6.58 cm). Other common presentations were vomiting ($n=12$, 29.27%), drowsiness ($n=7$, 17.07%), irritability ($n=10$, 24.40%), fever ($n=10$, 24.40%), sunset sign ($n=5$, 12.20%), convulsion ($n=4$, 9.76%), respiratory distress ($n=4$, 9.76%), bulging fontanel ($n=1$, 2.44%), and papilloedema ($n=2$, 4.88%). The distributions of presentations according to etiology were given in **Table 3**.

Out of 41 cases, 33 (80.49%) had improved outcomes, 7 (17.03%) stabilized, and 1(2.44%) had mortality during 6 to 24 months (mean: 17.85 ± 5.18) of follow-up. On follow-up, there were improvements in head circumference (range: 39–60 cm, mean: 46.15 ± 5.83 cm).

Discussion

Hydrocephalus is a common disorder of central nervous system in children. The soft compliant skull of infants has a significant ability to accommodate increased intracranial volume without creating an extreme elevation in intracranial pressure, simply by changing the head's size and shape. While increase in head size is only sign of hydrocephalus among most of the children below 2 years, older children may present with the classical triad of headache, vomiting and papilloedema. The development of shunt surgery has remarkably changed the outcome in these patients with better prospects of leading to a normal life. However, complications like infection and obstruction warranting shunt revisions continue to adversely affect the outcome.

Hydrocephalus is a common clinical condition leading to head enlargement in neonates.² It was a presentation in 30 (73.17%) cases and Dandy-Walker malformation in 4(9.76%) cases. Congenital aqueductal stenosis accounts for ~10% of all hydrocephalus cases in children.⁶ Aqueductal stenosis was the commonest cause of hydrocephalus in current study and

Table 3 Clinical characteristics

Clinical characteristics	HCP n (%)	DWM n (%)	SDC n (%)	AC n (%)	CS n (%)	TBM n (%)
Increased HC	28 (68.29%)	4 (9.76%)	2 (4.88%)	2 (4.88%)	1 (2.44%)	2 (4.88%)
Vomiting	8 (19.51%)	2 (4.88%)	1 (2.44%)	0	0	1 (2.44%)
Drowsiness	6 (14.63%)	0	0	0	0	1 (2.44%)
Irritability	9 (21.95%)	0	0	0	0	1 (2.44%)
Fever	8 (19.51%)	0	0	0	0	2 (4.88%)
Sunset sign	5 (12.20%)	0	0	0	0	0
Convulsions	4 (9.76%)	0	0	0	0	0
Respiratory distress	3 (7.32%)	0	0	0	0	1 (2.44%)
Bulging fontanel	18 (43.90%)	0	0	0	0	1 (2.44%)
Prominent scalp vein	13 (31.71%)	0	0	0	0	0
Abnormal head shape	0	0	0	0	1(2.44%)	0
Refusal to feed	0	1 (2.44%)	0	0	0	1 (2.44%)
Papilloedema	2 (4.88%)	0	0	0	0	0

Abbreviations: AC, arachnoid cyst; CS, craniosynostosis; DWM, Dandy-Walker malformation; HC, head circumference; HCP, hydrocephalus; SDC, subdural collection; TBM, tubercular meningitis.

accounted for 25(60.98%) cases of hydrocephalus. The disproportionately large number of aqueductal stenosis cases causing hydrocephalus in our study was because we had included cases only up to 6 months of age group in which congenital causes for hydrocephalus were more common and also due to small sample size of the study group.

Majority of the patients were clinically symptomatic on presentation to the hospital. Vomiting was present in 8 (19.51%) cases, irritability in 9 (21.95%), and fever in 10 (24.39%) cases. These were generally more commonly observed in hydrocephalic children. Increase in head circumference was the most common sign (100%) followed by tense anterior fontanel. Sunset sign, which probably represents upward gaze palsy due to compression of the tectal region of the brainstem,⁷ was very apparent in five (12.20%) cases. Study done by Schmidek HH et al⁸ had similar clinical presentation.

VPS surgeries were performed in majority of cases because of less success rate of endoscopic third ventriculostomy below 1 year of age group. Ventricular CSF shunting causes a list of complications, the most common is mechanical obstruction which was also observed in our study in five (12.20%) cases. This may occur at either one or both ends, but usually due to obstruction of ventricular catheter by entrapped choroid plexus tissue, intraventricular debris, or gliosis around the catheter tip. Proximal shunt obstruction is more common than the distal one.⁹ We had proximal obstruction in three of the five cases. Distal obstruction is due to accumulation of debris in the slit valve. We had two cases with peritoneal end blockage by tissue debris.

The second most common complication of these procedures is infection. This occurs in 5 to 10% of the cases and is usually the result of prior infection of CSF or infection introduced during surgery.¹⁰ Fortunately, we had not encountered this complication in our study. During a mean follow-up period, six (14.63%) cases required shunt revisions

due to shunt-related complications. One of these cases required multiple shunt revisions due to shunt blockage.

In Dandy-Walker malformation cases, after shunt surgery the cyst reduced in size and the bilateral cerebellar hemispheres grew markedly. We consider that treatment by CVPS shunt as adequate if the aqueduct is patent prior to surgery; even if hydrocephalus is a factor and that VPS placement is unnecessary.¹¹⁻¹⁴

Commonest treatment approach for benign SDC is observation with follow-up along with serial head circumference measurement, ultrasound, and computed tomography/magnetic resonance imaging. Most cases gradually resolve spontaneously, often within 8 to 9 months. Burr hole drainage may be needed in few patients who do not respond to conservative management.¹⁵ In our study, out of two cases one responded to conservative management very well. Our one case managed conservatively had failed; hence, burr hole drainage was done later. Subdural-peritoneal shunt (unilateral shunt) is usually adequate even for bilateral effusions.¹⁶ An extremely low-pressure system should be utilized. The general practice is to remove the shunt after 2 to 3 months of drainage. Subdural-peritoneal shunt was not needed in any of our patients.

The outcomes seem to depend on multiple factors. This appears to be related to earlier institution of therapy. In this subgroup these infants were in close observation since birth, which would mean that earlier therapeutic intervention has a significant effect on functional brain development. Early recognition and institution of therapy and close monitoring had led to improvement in head circumference in majority of our cases in this study. This will ultimately lead to the functional and neurological development in these cases.

What Is Next?

The treatment of children with hydrocephalus (one of the commonest cause of head enlargement) is far from perfect.

Hopefully, continued research in the area outlined in this manuscript will eventually pay dividends. Other areas of research not mentioned but of equal importance include fetal surgery, mathematical modeling of hydrocephalus (which in the future would be a powerful tool for the understanding and treatment of hydrocephalus as well as other conditions related to brain biomechanics), and tissue engineering (which aims to create custom designed living implantable devices for diverting CSF). Certainly, a significant start to improved treatment would be (a) the development of a shunt that would be truly physiological and (b) a simple test to determine the natural absorptive capacity of the patient.

Conclusion

The common neurosurgical causes of head enlargement in neonate and infants were hydrocephalus, Dandy-Walker malformation, and SDC. Shunt surgery was the most common form of management of these cases. Improvement in outcome of these children to normalcy or near normalcy appears to be possible with early detection and prompt institution of therapy. Serial and periodic follow-up program for diagnosing and treating complications and dysfunctions was the key for the successful outcomes in these patients.

Authors' Contributions

Dr. Sangh Mitra performed data collection. The remaining authors contributing in writing of the manuscript.

Funding

None.

Conflict of Interest

None declared.

References

- 1 Biran-Gol Y, Malinge G, Cohen H, et al. Developmental outcome of isolated fetal macrocephaly. *Ultrasound Obstet Gynecol* 2010; 36(02):147–153
- 2 DeMeyer W. Megalencephaly in children. Clinical syndromes, genetic patterns, and differential diagnosis from other causes of megalencephaly. *Neurology* 1972;22(06):634–643
- 3 Petersson S, Pedersen NL, Schalling M, Lavebratt C. Primary megalencephaly at birth and low intelligence level. *Neurology* 1999;53(06):1254–1259
- 4 Williams CA, Dagli A, Battaglia A. Genetic disorders associated with macrocephaly. *Am J Med Genet A* 2008;146A(15):2023–2037
- 5 Desch LW, Anderson SK, Snow JH. Relationship of head circumference to measures of school performance. *Clin Pediatr (Phila)* 1990;29(07):389–392
- 6 Dias MS. Myelomeningocele. In: Choux M, Di Rocco C, Hockley AD, eds. *Paediatric Neurosurgery*. London: Churchill Livingstone; 1999:253–69
- 7 Shallat RF, Pawl RP, Jerva MJ. Significance of upward gaze palsy (Parinaud's syndrome) in hydrocephalus due to shunt malfunction. *J Neurosurg* 1973;38(06):717–721
- 8 Schmidek HH, Auer LM, Kapp JP. The cerebral venous system. *Neurosurgery* 1985;17(04):663–678
- 9 Cozzens JW, Chandler JP. Increased risk of distal ventriculoperitoneal shunt obstruction associated with slit valves or distal slits in the peritoneal catheter. *J Neurosurg* 1997;87(05):682–686
- 10 Bayston R. Epidemiology, diagnosis, treatment, and prevention of cerebrospinal fluid shunt infections. *Neurosurg Clin N Am* 2001; 12(04):703–708, viii
- 11 Marinov M, Gabrovsky S, Undjian S. The Dandy-Walker syndrome: diagnostic and surgical considerations. *Br J Neurosurg* 1991;5 (05):475–483
- 12 Bindal AK, Storrs BB, McLone DG. Management of the Dandy-Walker syndrome. *Pediatr Neurosurg* 1990–1991;16(03):163–169
- 13 Domingo Z, Peter J. Midline developmental abnormalities of the posterior fossa: correlation of classification with outcome. *Pediatr Neurosurg* 1996;24(03):111–118
- 14 Kalidasan V, Carroll T, Allcutt D, Fitzgerald RJ. The Dandy-Walker syndrome—a 10-year experience of its management and outcome. *Eur J Pediatr Surg* 1995;5(Suppl 1):16–18
- 15 Greenberg MS. *Handbook of Neurosurgery*. 7th edition. New York: Thieme Medical Publishers; 2010:904–905
- 16 Aoki N. Chronic subdural hematoma in infancy. Clinical analysis of 30 cases in the CT era. *J Neurosurg* 1990;73(02):201–205