of the mandible. The common presence of local irritating factors in the anterior region of the mandible could explain this finding where the local irritants are highly involved in pathogenesis of oral PGs.[3,9,10]

Based on the results of this study we concluded that the highest recurrence rate of oral giant PG occurs in mandibular lesions of the third and fourth decades of life with a female predilection. So, these lesions must be carefully excised with removal of the involved teeth if they were loose. Also, the patients should be observed at least for 1 year after surgery because of the high recurrence rate.

REFERENCES

1. Gordón-Núñez MA, de Vasconcelos Carvalho M, Benevenuto TG, et al. Oral pyogenic granuloma: a retrospective analysis of 293 cases in a Brazilian population. *J Oral Maxillofac Surg* 2010;68:2185–2188
2. Krishnapillai R, Punnoose K, Angadi PV, et al. Oral pyogenic granuloma—a review of 215 cases in a South Indian Teaching Hospital, Karnataka, over a period of 20 years. *Oral Maxillofac Surg* 2012;16:305–309
3. da Silva AD, Silva CA, de Camargo Moraes P, et al. Recurrent oral pyogenic granuloma in port-wine stain. *J Craniofac Surg* 2011;22:2356–2358
4. Saravana GH. Oral pyogenic granuloma: a review of 137 cases. *Br J Oral Maxillofac Surg* 2009;47:318–319
5. Fekrazad R, Nokhbatolfoghahaei H, Khoei F, et al. Pyogenic granuloma: surgical treatment with Er:YAG laser. *J Lasers Med Sci* 2014;5:199–205
6. Ximenes M, Triches TC, Cardoso M, et al. Pyogenic granuloma on the tongue: a pediatric case report. *Gen Dent* 2013;61:27–29
7. Sridhar R, Wanjari S, Kanteshwari IK. Interrelationship between pyogenic granuloma and peripheral ossifying fibroma: a case report. *J Dent Hyg* 2012;86:179–184
8. Al-Khateeb T, Ababneh K. Oral pyogenic granuloma in Jordanians: a retrospective analysis of 108 cases. *J Oral Maxillofac Surg* 2003;61:1285–1288
9. Asnaashari M, Mehdipour M, MoradiAbbasabadi F, et al. Expedited removal of pyogenic granuloma by diode laser in a pediatric patient. *J Lasers Med Sci* 2015;6:40–44
10. Kamal R, Dahiya P, Puri A. Oral pyogenic granuloma: various concepts of etiopathogenesis. *J Oral Maxillofac Pathol* 2012;16:79–82
11. Punde PA, Malik SA, Malik NA, et al. Idiopathic huge pyogenic granuloma in young and old: an unusually large lesion in two cases. *J Oral Maxillofac Pathol* 2013;17:463–466
12. Gomes SR, Shakir QJ, Thaker PV, et al. Pyogenic granuloma of the gingiva: a misnomer?—A case report and review of literature. *J Indian Soc Periodontol* 2013;17:514–519
13. Mahabob N, Kumar S, Raja S. Palatal pyogenic granulomaa. *J Pharm Bioallied Sci* 201;5 (suppl 2):S179–S181
14. Singh RK, Kaushal A, Kumar R, et al. Profusely bleeding oral pyogenic granuloma in a teenage girl. *BMJ Case Rep* 2013. doi: 10.1136/bcr-2013-008583
15. Asnaashari M, Bigom-Taheri J, Mehdipoor M, et al. Posthaste outgrow of lip pyogenic granuloma after diode laser removal. *J Lasers Med Sci* 2014;5:92–95
16. Brunet-LLobet L, Miranda-Rius J, Lahor-Soler E, et al. A gray-purple mass on the floor of the mouth: gigantic mucogingival pyogenic granuloma in a teenage patient. *Open Dent J* 2014;8:125–128
17. Lindenmüller IH, Noll P, Mameghani T, et al. CO2 laser-assisted treatment of a giant pyogenic granuloma of the gingiva. *Int J Dent Hyg* 2010;8:249–252
18. Frumkin N, Nashef R, Shapira L, et al. Nonsurgical treatment of recurrent gingival pyogenic granuloma: a case report. *Quintessence Int* 2015;46:539–544
19. Wine Lee L, Goff KL, Lam JM, et al. Treatment of pediatric pyogenic granulomas using β-adrenergic receptor antagonists. *Ediatr Dermatol* 2014;31:203–207
20. Ichimiya M, Yoshikawa Y, Hamamoto Y, et al. Successful treatment of pyogenic granuloma with injection of absolute ethanol. *J Dermatol* 2004;31:342–344
21. Matsumoto K, Nakanishi H, Seike T, et al. Treatment of pyogenic granuloma with a sclerosing agent. *Dermatol Surg* 2001;27: 521–523
22. Ghalayani P, Hajisadeghi S, Babadi F. Extragingival pyogenic granuloma associated with medication: report of an unusual case. *Dent Res J (Isfahan)* 2014;11:400–404
23. Samatha Y, Reddy TH, Jyothirrmai K, et al. Management of oral pyogenic granuloma with sodium tetra decyl sulphate. A case series. *N Y State Dent J* 2013;79:55–57

Management of Craniosynostosis at an Advanced Age: Controversies, Clinical Findings, and Surgical Treatment

Rajiv J. Iyengar, BS, Petra M. Klinge, MD, PhD,† Wendy S. Chen, MD, PhD,‡ Jerrold L. Boxerman, MD, PhD,§ Stephen R. Sullivan, MD, MPH,|| and Helena O. Taylor, MD, PhD||*

Background: The natural history of unrepaired craniosynostosis is not well defined. Delayed surgical intervention carries greater risk of postoperative complications and its functional benefits for older patients are poorly characterized. The authors reviewed patients in whom children presented beyond 1 year of age to better understand the natural history of craniosynostosis, and the risk–benefit relationship for delayed reconstruction.

Methods: After institutional IRB approval the authors conducted a retrospective review of patients who presented after 1 year of age with craniosynostosis. Type of craniosynostosis, age at evaluation, medical history, surgical findings, developmental abnormalities, ophthalmologic findings, and clinical course were reviewed.

Results: Ten patients with delayed presentation for craniosynostosis were identified. The mean age at presentation was 6.8

From the *Warren Alpert Medical School of Brown University; †Department of Neurosurgery; ‡Division of Ophthalmology, Warren Alpert Medical School of Brown University, Rhode Island Hospital and Hasbro Children's Hospital; §Department of Diagnostic Imaging, Rhode Island Hospital and Warren Alpert Medical School of Brown University; and ||Division of Plastic Surgery, Mount Auburn Hospital, Cambridge, MA.

Received October 27, 2015.
Accepted for publication March 4, 2016.

Address correspondence and reprint requests to Helena O. Taylor, MD, PhD, Division of Plastic Surgery, Mount Auburn Hospital, 300 Mt Auburn Street, Suite 304, Cambridge MA 02138;
E-mail: htaylor@mah.harvard.edu

The authors report no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0000000000002725

years ± 4.2 years (range, 3–17 years). Seven of 10 patients presented with developmental delay. Five patients presented with debilitating headaches. Five patients presented with comorbid Chiari malformations, 3 of whom required surgical decompression. Two patients had papilledema. Four patients underwent intracranial pressure monitoring, with elevated pressures found in 3 patients. Six patients underwent delayed cranial vault remodeling. There were no peri- or postoperative complications, including infection or residual bony defects, in those undergoing delayed operation.

Conclusions: Children who present in a delayed fashion with unrepaired craniosynostosis have high rates of debilitating headaches, developmental delays, head shape anomalies, and Chiari malformation. Five patients reporting preoperative headaches noted subjective improvements in headaches following delayed operation. Cranial reconstruction can be safely performed at an older age and is appropriate to consider in carefully selected patients for aesthetic and/or functional concerns.

Key Words: Advanced age, cerebrospinal fluid flow, Chiari malformation, cranial expansion, cranial remodeling, cranial vault, cranioplasty, craniosynostosis, delayed, intracranial pressure, MRI

The current standard of care for craniosynostosis involves cranial vault remodeling within the first year of life.[1] While there is much debate regarding exact timing and type of procedure, few advocate delaying reconstructions beyond 1 year of age.

The phenotypic effects of cranial remodeling are readily apparent, but the effects on intracranial pressure, neurodevelopment, vision, and risk of Chiari malformation are controversial.[2] Intracranial pressure is particularly difficult to interpret given the lack of normative data in children,[3,4] Neurodevelopment in craniosynostosis is also a much-debated topic considering that the majority of early studies did not establish control groups to allow for robust analysis.[5] Kapp–Simon et al recently conducted a more rigorous study and found that children with single-suture craniosynostosis had adjusted IQ scores of 2.5 to 4 points lower than unaffected children,[6] but it is hard to discern if this is clinically significant. The lack of clear consensus data on the benefits of cranial vault reconstruction on intracranial pressure (ICP) and development makes it challenging to counsel families regarding the benefits of cranial remodeling and expansion.[7] This is especially difficult in patients with minimal skull deformity, or in older patients who may have completed much of their brain and calvarial growth. As cranial vault reconstructions are almost always performed once the diagnosis is made, it is rare to encounter older children in whom the natural history of unoperated craniosynostosis can be assessed.

In this study, we describe a cohort of patients with a delayed presentation of nonsyndromic craniosynostosis. We review their clinical and radiological findings to characterize the natural history of their pathology. We also review the subset of patients who underwent cranial reconstruction in a delayed fashion. The perioperative courses of these patients were compared to 7 sequential patients who were treated in infancy for their craniosynostosis, to assess the relative risks and benefits of reconstruction in these older children. Currently acceptable approaches include open cranial vault remodeling, strip craniectomy with adjuvant helmet therapy,[8] and strip craniectomy with springs.[9]

METHODS

After IRB approval, the electronic medical records at our institution were used to identify consecutive patients with delayed diagnosis and treatment of craniosynostosis over a 7-year period between January 2008 and February 2015. Inclusion criteria were age greater than 2 years at the time of initial evaluation, fusion of at least 1 suture, and adequate imaging studies including computed tomography scans. Several of the subjects underwent magnetic resonance imaging (MRI) scanning with additional cine cerebrospinal fluid (CSF) flow studies to help characterize changes in CSF flow at the foramen magnum. Exclusion criteria were syndromic diagnosis or previous cranial vault remodeling with craniectomy. We recorded type of craniosynostosis, age at evaluation, symptoms, past medical history, surgical management, developmental abnormalities, ophthalmologic findings, and clinical course. For those who underwent reconstruction, surgical duration, blood transfusion volume, and hospital length of stay were recorded. These patients were matched to 6 sequential patients in the same time period who underwent cranial vault reconstruction in infancy for comparison of these perioperative measures of morbidity.

All children were evaluated by a multidisciplinary team including a craniofacial plastic surgeon, neurosurgeon, ophthalmologist, neuroradiologist, developmental specialist, and medical geneticist. For delayed patients, our typical treatment algorithm can be seen in Figure 1. While head shape concerns and functional concerns are depicted as a strict dichotomy in this figure, the reality is that many patients may have both head shape and potential ICP concerns. Head shape anomaly was graded as follows: normal/mild (+), moderate (++), or severe (+++). The presence of Chiari malformations and pulsatility of CSF on cine phase-contrast MRI were assessed by a neuroradiologist. Specifically, the extent of cerebellar tonsil ectopia below the basion–opisthion line was measured on sagittal T1-weighted images of the brain. Additionally, we qualitatively evaluated the phase images from the sagittal cine CSF flow series obtained at midline using velocity encoding (venc) values of 5 and 10 cm/s for any evidence of reduced "phasicity" of CSF with the cardiac cycle at the foramen magnum, including ventral and dorsal to cervicomedullary junction and about the cerebellar tonsil tips. In the absence of established treatment recommendations for these patients, our multidisciplinary team made a concerted effort to include the family in shared decision making. The priorities and concerns of the families are weighted heavily in the treatment choices. In borderline patients, either preoperative intraparenchymal ICP monitoring was performed for 24 to 48 hours or epidural ICP monitoring intraoperatively, at the time of reconstruction, through a frontal burr hole with recording of baseline ICP for 10 minutes.[10] Intracranial pressure was considered abnormal in the 24 to 48 hours monitoring period if A-wave or frequent B-wave activity was present as described previously[11] or if baseline measurement intraoperatively exceeded 15 mm Hg.[10]

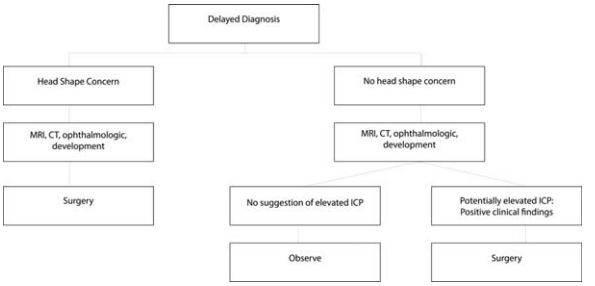

FIGURE 1. Current algorithm for management of craniosynostosis identified in children greater than 1 year of age.

TABLE 1. Patient Demographic Data

Cohort n = 10	
Sex (male:female)	7:3
Mean age at presentation	6.8 ± 4.2 yrs
Type of craniosynostosis	5 sagittal
	2 sagittal + bilateral lambdoid
	2 pan suture
	1 metopic
Syndromic patients	None

TABLE 3. Summary of Clinical Findings

Cohort	n = 10
Chiari malformation	5 (50%)
Cine MRI (diminished CSF flow)	4 (40%)
Headaches	5 (50%)
Documented ICP elevation	3 (30%)
Ophthalmologic findings	2 (20%)
Developmental delay	7 (70%)

ICP, intracranial pressure; MRI, magnetic resonance imaging.

RESULTS

We identified 10 patients with a delayed diagnosis of craniosynostosis. One patient who had undergone extracranial shaving of her trigonocephaly as an infant was also included (patient 2). Syndromic patients were intentionally excluded to minimize confounding factors on development and ICP. Table 1 summarizes the demographics of this population.

Preoperative diagnoses, surgical history, symptoms, and comorbidities are presented in Table 2 and a summary of clinical findings is presented in Table 3. Seven patients (70%) exhibited either moderate or severe head shape anomalies, but some had relatively normal head shape; superior views of all 3D computed tomography reconstructions are shown in Figure 2. Developmental delay was the most common comorbidity, occurring in 7 patients (70%). Five patients (50%) had Chiari malformations, defined as a >5 mm cerebellar tonsillar ectopia below the basion–opisthion line on MRI, and 4 of these had associated diminished CSF pulsatility at the foramen magnum as detected by MRI cine flow studies. The spectrum of MRI findings is shown in Figure 3. Five patients (50%) presented with chronic nonspecific, nonfocal headaches with concern for increased intracranial pressure. Four (40%) underwent ICP measurements either at the time of surgery (epidural monitoring) or prior to surgery (intraparenchymal monitoring), and 3 of these patients (30% of total, and 75% of those measured) had documented elevations in ICP. Two patients (20%) exhibited papilledema.

The majority (60%) of patients in this cohort underwent cranial vault remodeling. Figure 4 outlines the preoperative clinical findings of 6 patients who underwent delayed operation. The family of 1 patient (patient 1) with severe delays was reluctant to proceed with cranial remodeling. One patient (patient 2) had her trigonocephaly shaved down in infancy and underwent Chiari decompression for a symptomatic Chiari malformation, but has not had open cranial vault remodeling. One patient was evaluated at our institution, but her family elected to undergo Chiari decompression at another institution and forego cranial remodeling (patient 4).

The comparative perioperative data (median estimated blood loss, operative time, and hospital stay) between reconstructions on infants versus older children is shown in Table 4. Delayed surgery required greater operative time (401.5 ± 42.9 minutes versus 341.0 ± 39.6 minutes, $P = 0.00135$), but differences between the 2 groups for estimated blood loss (650.0 ± 232.6 cc versus

TABLE 2. Clinical Data and Findings

Patient	Sex	Age at Operation	Fused Suture (s)	Surgery	Cine	Chiari	HA	DD	PE	ICP (mm Hg)	Head Shape	Percentage Increase in Intracranial Volume
1	M	6	Pan suture	None	DF	+	+	+	−	N/A	++	N/A
2	F	6	Metopic	Shaving metopic ridge, prior Chiari decompression	DF	+	+	−	−	N/A	++	N/A
3	M	11	Sagittal	Subtotal CVR	Normal	−	−	+	−	Epidural pressures 7–10 (normal)	++	+14%
4	F	4	Sagittal	Chiari decompression at Yale, No CVR	DF	+	−	+	−	N/A	++	N/A
5	M	5	Sagittal	Subtotal CVR	Normal	−	−	+	−	N/A	+++	Unavailable
6	F	3	Sagittal	Subtotal CVR	Normal	−	+	+	−	Epidural pressures 17–22	+	+3%
7	M	17	Pan Suture	Prior Chari decompression, total CVR	DF	+	+	+	+	Intraparenchymal pressures >20	+	+18%
8	M	5	Sagittal, bilateral lambdoid	Subtotal CVR	Normal	−	−	−	−	N/A	+++	+15%
9	M	4	Sagittal, bilateral lambdoid	Subtotal CVR	Normal	−	−	+	−	N/A	+	Unavailable
10	M	7	Sagittal	Subtotal CVR	Normal	+	+	−	+	Intraparenchymal pressures >40	++	+12%

CVR, cranial vault remodeling.

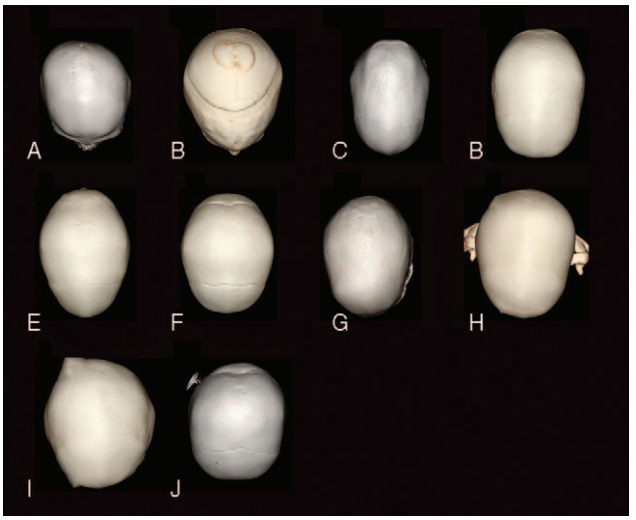

FIGURE 2. (A–J) Three-dimensional computed tomography calvarial reconstructions of patients 1 to 10 (as described in Table 2) presented sequentially.

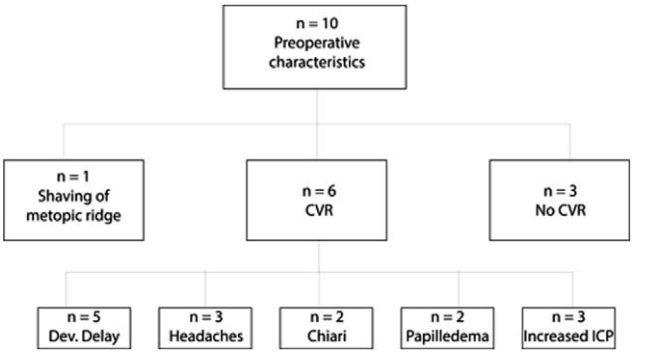

FIGURE 4. Preoperative characteristics of 6 patients who underwent delayed cranial vault operations. CVR, cranial vault remodeling; Dev. Delay, developmental delay; ICP, intracranial pressure.

550.0 ± 173.5 cc, $P = 0.499$) and duration of hospital stay (median = 4 days for both groups, $P = 0.764$) did not achieve statistical significance. One patient (patient 10) was excluded from the hospital stay analysis, as he remained in the hospital for shunt related concerns, not as a result of his cranial vault surgery. The mean percentage increase in intracranial volume following the cranial vault remodeling was 12% (range, 3–18%) in the 5 patients for whom we have pre- and postoperative volumetric data (Table 2). There were no complications (eg, return to OR, prolonged intubation) in the cohort of older children, and 1 return to the OR for washout in the infant controls for suspicion for infection.

DISCUSSION

Craniosynostosis is most often recognized shortly after birth due to the presence of skull deformities corresponding to the involved sutures.[3] Parameters of care for craniosynostosis have been reviewed and summarized by a multidisciplinary panel of craniofacial experts,[1] but there remain many areas of ambiguity. While the optimal timing and technique for reconstruction are debated, surgical intervention for nonsyndromic craniosynostosis is usually performed within the first year of life.[12,13] We present an unusual subset of patients who were older than 1 year of age at the time of presentation. Counseling these patients and their families is a challenge given the paucity of data on intervention in this age group. These patients provide useful insight into the natural history of unoperated craniosynostosis, as well as the risks and benefits of delayed reconstruction.

Older pediatric patients may require a more extensive operation and have decreased calvarial regenerative potential, increasing the risk for residual skull defects.[14] The development of a pneumatized frontal sinus may increase risk of infection. The exponential growth phase of the young brain is largely complete after 3 years of age,[15] which further heightens the need for robust data on the functional utility of delayed cranial vault remodeling. These factors complicate the surgical decision-making process, necessitating a critical assessment of surgical risk versus benefit.

Normalization of head shape is an accepted indication for cranial vault remodeling. Marchac et al[16] published a series of 13 adult patients with unoperated craniosynostosis, who presented in a delayed fashion with aesthetic concerns. They concluded that cranial remodeling was beneficial from an aesthetic standpoint in most patients, and could be safely performed even in adulthood. Developmental and functional concerns were not rigorously evaluated in their analysis.

Our findings also indicate that delayed surgery is safe to perform. Median estimated blood loss, median operative time, and median hospital stay were comparable between the children undergoing delayed operations and those receiving operations at infancy (Table 4). There were no complications or adverse events associated with delayed surgical intervention, which is reassuring to the surgeon and anxious families making difficult decisions about management. The only complication in the subjects or controls was a surgical site infection in a 1-year old who underwent timely cranial vault remodeling for metopic synostosis. This was treated with washout in the OR and a short course of antibiotics.

Internal distraction osteogenesis has produced aesthetic and functional improvement in older children with scaphocephaly

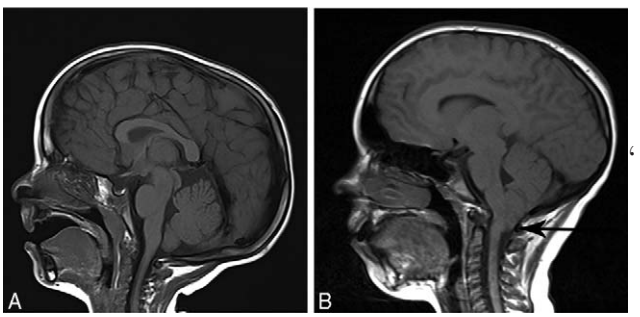

FIGURE 3. (A) MRI scan from patient 5 demonstrating a normal posterior fossa. (B) MRI scan from patient 7 revealing a Chiari malformation (black arrow points to area of tonsillar herniation). MRI, magnetic resonance imaging.

TABLE 4. Comparison of Surgical Parameters Between Delayed and Timely Cranial Vault Reconstructions

	Delayed cranial vault n = 6	Cranial vault in infancy n = 6	*P* (2-tailed *t*-test)
Mean estimated blood loss (EBL)	650 ± 233 cc	550 ± 174 cc	0.499
Median operative time	402 ± 43 min	341 ± 40 min	0.00135
Median hospital stay	4 days	4 days	

secondary to sagittal suture synostosis and symptoms of increased intracranial pressure.[17] These authors argue that distraction is an appropriate therapeutic intervention for children with symptomatic scaphocephaly, and that operative time is shorter and there are few residual bony defects following the procedure. However, distraction requires multiple operations and offers only 2-dimensional correction of cephalocranial disproportion. Several patients in our cohort presented with multi- or pan-suture synostosis, for which full cranial vault remodeling enables more global correction and expansion.

Those who believe that cranial vault remodeling is largely an aesthetic procedure often cite this lack of a discernable correlation between size and pressure metrics.[18] Gault et al[19] found that volume measurement alone did not reliably predict increased intracranial pressure in children with craniosynostosis. Factors other than cranial volume such as abnormal venous drainage, airway obstruction, and abnormalities in cerebrospinal fluid dynamics have been proposed to explain the persistence of elevated intracranial pressure following cranial vault remodeling in certain patients.[20] Intracranial volume is however 1 quantitative measure of the impact of the operation on cranial morphology. We document a 12% mean (range, 3–18%) increase in the 5 patients who underwent cranial vault remodeling and had analyzable volumetric data. Following the operation all of these patients experienced improvement in symptoms.

While the presence of elevated intracranial pressure is often discussed as a quantitative factor linked to neurodevelopmental outcome, this correlation has yet to be proven,[21,22] Minns[23] established rough guidelines for upper normal limits of intracranial pressure in patient populations of various ages, ranging from 3.5 mm Hg in neonates to 15.3 mm Hg in adolescents/adults. Though these guidelines exist, the lack of scientific consensus on normal intracranial pressure values in children complicates the interpretation of this data.[24] Intraparenchymal intracranial pressure monitoring over 24 to 48 hours is the gold standard for the measurement of intracranial pressure; but, it requires an invasive surgical approach, and prolonged hospital stay.[25] There are currently no reliable radiographic surrogates employed in routine clinical practice to screen for elevated ICP.[26] Anecdotally, in developing countries some children with craniosynostosis are treated with shunting alone, but there is little in the literature to demonstrate the efficacy of this approach. Shunting may temper elevations in pressure, but does not normalize CSF dynamics or cranial morphology. Data from the hydrocephalus population also demonstrates that the morbidity of shunting is significant.[27]

The presence of papilledema has long been used as a clinical marker of increased intracranial pressure, but the low sensitivity in the younger pediatric population can complicate reliable implementation.[28] Our cohort corroborated that papilledema in craniosynostosis is an imperfect indicator of elevated ICP; only 2 of the 3 patients with documented ICP elevation in our study exhibited ophthalmologic findings consistent with papilledema. While some centers are routinely measuring intracranial pressures invasively prior to recommending surgery,[29] this has not been established as a standard treatment algorithm in the United States.

Intracranial pressure tracings performed on 4 of our patients (2 epidural intraoperative and 2 intraparenchymal preoperative) revealed prolonged elevations in pressure readings from 3 patients (30% of total population and 75% of those measured), nearing or exceeding 20 mm Hg (Table 2). On the ICP tracing, the presence of A waves revealed acute elevations of ICP and B waves demonstrated volatile ICP, consistent with a pathological state and decompensation of the normal autoregulatory mechanisms.[25] Of note, the craniofacial group from Oxford published similar findings from a cohort of older children with uncorrected nonsyndromic unicoronal synostosis in which they found elevated ICP in 5 of 7 children.[30]

The craniofacial group at the University of Washington demonstrated an improvement in symptoms and/or signs of elevated intracranial pressure in 17 older children with delayed cranial vault expansion.[31] Seven of these patients had never undergone any surgical intervention for craniosynostosis, while 10 had been operated on previously. Overall, 12 of 14 patients experienced a remission of headaches, 9 patients experienced improvement in nausea and vomiting, and 4 patients experienced resolution of papilledema following the cranial vault. The authors did not, however, include an analysis of older children with craniosynostosis who lacked symptoms of elevated ICP, reflective of the referral bias toward symptomatic children. It is therefore difficult to draw conclusions about the prevalence of elevated ICP in children with unoperated craniosynostosis from this study.

We also found considerable qualitative postoperative functional improvement in several patients. Patient 7, a 17-year-old boy previously described in detail,[11] presented with a relatively normal head shape and mild microcephaly in the setting of pan-suture synostosis, developmental delay, symptoms of ICP elevation, and a Chiari malformation (Table 2). He experienced lasting relief from debilitating headaches after undergoing cranial vault remodeling.

By contrast, patient 8 from our cohort underwent cranial vault remodeling to correct a severe head shape anomaly without any associated symptoms of ICP increase or Chiari malformation despite a diagnosis of multisuture synostosis. Patient 5 also presented with a severe head shape anomaly secondary to his sagittal synostosis but did not exhibit any neurological symptoms other than mild developmental delay. Patient 6, however, had a relatively normal head shape but exhibited developmental deficits and elevated ICP (Table 2, Fig. 2). Thus, the degree of cranial dysmorphology appears to be a poor predictor of underlying neurological symptoms in older patients with uncorrected craniosynostosis, consistent with previous reports in the literature.[30,32]

The effect of surgical intervention on developmental trajectory remains difficult to measure, in part because of the lack of a consistent metric for development. Kapp-Simon et al[33] suggest a complex relationship between normalization of head shape and development perhaps because of abnormalities in the maturation of the brain itself. Correction of cranial vault dysmorphology can improve the shape of the brain, but the functional implications of this are unclear.[34] Subtle deficits in children with craniosynostosis have been well documented; reading and spelling ability was deficient in 50% of children with sagittal suture synostosis following nuanced analysis.[35] A striking 7 of 10 of our patients presented with developmental delay based on evaluations from our institutional specialists, including 5 of 6 with single suture synostosis and 2 of 4 of the multisuture craniosynostosis patients (Tables 2 and 3). These findings in our delayed cohort demonstrate the persistence of these neurocognitive deficits through early childhood and adolescence. The nature of this retrospective study cannot, however, control for confounders, such as familial resources and education, which could also account for delayed presentation. The craniofacial group at Yale has recently contributed an impressive body of work examining development in children with sagittal synostosis, and the impact of surgical timing,[36] but additional prospective studies are needed.

The comorbidity of craniosynostosis and Chiari malformations has been documented extensively in syndromic patients of multisuture craniosynostosis, and lambdoid suture synostosis (25), but its coincidence with other single suture synostoses is less well characterized.[37] Magnetic resonance imaging is not routinely performed for nonsyndromic patients, so the potential exists for missed diagnoses of Chiari malformations. In this series we found Chiari malformations to be quite common, present in 5 of 10 (50%) of our patients. Four of 5 of our patients with Chiari malformations exhibited qualitatively diminished CSF pulsatility at the foramen magnum on Cine MRI,[38] adding further support to the obstructive effects of a Chiari malformation on CSF circulation. Whether these

hindbrain herniations occur concomitantly, or in a delayed fashion as a result of the untreated craniosynostosis remains unclear. No algorithm currently exists to guide surgeons as to whether to decompress the Chiari before, during, or after cranial vault remodeling, but it is our impression that mild Chiari malformations may resolve with posterior vault expansion without formal Chiari decompression. The frequency of Chiari malformations in this series calls for greater clinical monitoring in these patients.

In summary, we believe that the benefits outweigh the risks for delayed cranial vault remodeling in carefully selected older patients with unoperated craniosynostosis presenting with headache, increased ICP, and/or moderate-to-severe head shape anomaly. Estimated blood loss, operative time, and length of hospital stay have been cited as potential concerns in delayed open operations.[8] In our cohort, delayed operations required slightly greater operative time, which was expected given the higher calvarial bone density in older children; blood loss and hospital stay data, however, did not demonstrate statistically significant differences between the 2 groups (Table 4). The improvement in headaches experienced by patients and lack of major postoperative complications further indicate a good safety profile for delayed operation. It is therefore reasonable and safe to consider cranial vault remodeling in older pediatric patients, who eluded timely diagnosis and treatment of craniosynostosis at infancy.

CONCLUSIONS

While the debate rages over the functional utility of cranial vault remodeling for nonsyndromic craniosynostosis, this series suggests a high rate of morbidity in neglected patients. Seven of 10 children had documented developmental delay, 5 of 10 had Chiari malformations, and 5 of 10 had debilitating headaches. Three presented with documented ICP elevation. This series suggests that delaying cranial vault reconstruction for craniosynostosis is associated with a high incidence of developmental delay, headaches, and coincident Chiari malformations. Delayed reconstruction and cranial vault expansion can be performed for these older patients with low morbidity but added operative time. While the indications to operate on craniosynostosis in infancy are well established, we can ameliorate aesthetic and functional concerns with delayed cranial vault remodeling in older pediatric patients. The significant morbidity seen in this cohort calls for increased clinical vigilance for signs and symptoms of elevated ICP, Chiari malformations, and developmental delay in these older children with untreated craniosynostosis.

REFERENCES

1. Warren SM, Proctor MR, Bartlett SP, et al. Parameters of care for craniosynostosis: craniofacial and neurologic surgery perspectives. *Plast Reconstr Surg* 2012;129:731–737
2. Chim H, Gosain AK. An evidence-based approach to craniosynostosis. *Plast Reconstr Surg* 2011;127:910–917
3. Marchac D, Renier D. Treatment of craniosynostosis in infancy. *Clin Plast Surg* 1987;14:61–72
4. Thompson DN, Malcolm GP, Jones BM, et al. Intracranial pressure in single-suture craniosynostosis. *Pediatr Neurosurg* 1995;22:235–240
5. Speltz ML, Kapp-Simon KA, Cunningham M, et al. Single-suture craniosynostosis: a review of neurobehavioral research and theory. *J Pediatr Psychol* 2004;29:651–668
6. Speltz ML, Collett BR, Wallace ER, et al. Intellectual and academic functioning of school-age children with single-suture craniosynostosis. *Pediatrics* 2015;135:e615–e623
7. Persing JA, Mayes L, Bridgett D, et al. Commentary: the effects of whole vault cranioplasty versus strip craniectomy on long-term neuropsychological outcomes in sagittal craniosynostosis. *Plast Reconstr Surg* 2015;136:115e–117e
8. Jimenez DF, Barone CM, Cartwright CC, et al. Early management of craniosynostosis using endoscopic-assisted strip craniectomies and cranial orthotic molding therapy. *Pediatrics* 2002;110 (1 Pt 1):97–104
9. Guimaraes-Ferreira J, Gewalli F, David L, et al. Spring-mediated cranioplasty compared with the modified pi-plasty for sagittal synostosis. *Scand J Plast Reconstr Surg Hand Surg* 2003;37:208–215
10. Tans JT, Poortvliet DC. Intracranial volume-pressure relationship in man. Part 1: calculation of the pressure-volume index. *J Neurosurg* 1982;56:524–528
11. Iyengar RJ, Klinge PM, Chen WS, et al. Management of craniosynostosis at an advanced age: clinical findings and interdisciplinary treatment in a 17 year-old with pan-suture synostosis. *Interdisciplinary Neurosurg Adv Tech Case Manage* 2015;2:61–64
12. Marchac D, Renier D, Broumand S. Timing of treatment for craniosynostosis and facio-craniosynostosis: a 20-year experience. *Br J Plast Surg* 1994;47:211–222
13. Polley JW, Charbel FT, Kim D, et al. Nonsyndromal craniosynostosis: longitudinal outcome following cranio-orbital reconstruction in infancy. *Plast Reconstr Surg* 1998;102:619–628
14. Rottgers SA, Kim PD, Kumar AR, et al. Cranial vault remodeling for sagittal craniosynostosis in older children. *Neurosurg Focus* 2011;31:E3
15. Chugani HT, Phelps ME, Mazziotta JC. Positron emission tomography study of human brain functional development. *Ann Neurol* 1987;22:487–497
16. Marchac D, Renier D, Arnaud E. Unoperated craniosynostosis patients: correction in adulthood. *Plast Reconstr Surg* 2008;122:1827–1838
17. Lao WW, Denny AD. Internal distraction osteogenesis to correct symptomatic cephalocranial disproportion. *Plast Reconstr Surg* 2010;126:1677–1688
18. Thompson DN, Jones BM, Harkness W, et al. Consequences of cranial vault expansion surgery for craniosynostosis. *Pediatr Neurosurg* 1997;26:296–303
19. Gault DT, Renier D, Marchac D, et al. Intracranial pressure and intracranial volume in children with craniosynostosis. *Plast Reconstr Surg* 1992;90:377–381
20. Hayward R. Venous hypertension and craniosynostosis. *Childs Nerv Syst* 2005;21:880–888
21. Arnaud E, Renier D, Marchac D. Prognosis for mental function in scaphocephaly. *J Neurosurg* 1995;83:476–479
22. Gewalli F, Guimaraes-Ferreira JP, Sahlin P, et al. Mental development after modified pi procedure: dynamic cranioplasty for sagittal synostosis. *Ann Plast Surg* 2001;46:415–420
23. Minns RA. Intracranial pressure monitoring. *Arch Dis Child* 1984;59:486–488
24. Eide PK, Helseth E, Due-Tonnessen B, et al. Assessment of continuous intracranial pressure recordings in childhood craniosynostosis. *Pediatr Neurosurg* 2002;37:310–320
25. Czosnyka M, Pickard JD. Monitoring and interpretation of intracranial pressure. *J Neurol Neurosurg Psychiatry* 2004;75:813–821
26. Tamburrini G, Caldarelli M, Massimi L, et al. Intracranial pressure monitoring in children with single suture and complex craniosynostosis: a review. *Childs Nerv Syst* 2005;21:913–921
27. Paulsen AH, Lundar T, Lindegaard KF. Pediatric hydrocephalus: 40-year outcomes in 128 hydrocephalic patients treated with shunts during childhood. Assessment of surgical outcome, work participation, and health-related quality of life. *J Neurosurg Pediatr* 2015;16:633–641
28. Tuite GF, Chong WK, Evanson J, et al. The effectiveness of papilledema as an indicator of raised intracranial pressure in children with craniosynostosis. *Neurosurgery* 1996;38:272–278
29. Wilkie AO, Wall SA. Craniosynostosis: novel insights into pathogenesis and treatment. *Curr Opin Neurol* 1996;9:146
30. Eley KA, Johnson D, Wilkie AO, et al. Raised intracranial pressure is frequent in untreated nonsyndromic unicoronal synostosis and does not correlate with severity of phenotypic features. *Plast Reconstr Surg* 2012;130:690e–697e
31. Scott JR, Isom CN, Gruss JS, et al. Symptom outcomes following cranial vault expansion for craniosynostosis in children older than 2 years. *Plast Reconstr Surg* 2009;123:289–297
32. Morritt DG, Yeh FJ, Wall SA, et al. Management of isolated sagittal synostosis in the absence of scaphocephaly: a series of eight cases. *Plast Reconstr Surg* 2010;126:572–580

33. Kapp-Simon KA, Speltz ML, Cunningham ML, et al. Neurodevelopment of children with single suture craniosynostosis: a review. *Childs Nerv Syst* 2007;23:269–281
34. Aldridge K, Kane AA, Marsh JL, et al. Relationship of brain and skull in pre- and postoperative sagittal synostosis. *J Anat* 2005;206:373–385
35. Magge SN, Westerveld M, Pruzinsky T, et al. Long-term neuropsychological effects of sagittal craniosynostosis on child development. *J Craniofac Surg* 2002;13:99–104
36. Patel A, Yang JF, Hashim PW, et al. The impact of age at surgery on long-term neuropsychological outcomes in sagittal craniosynostosis. *Plast Reconstr Surg* 2014;134:608e–617e
37. Cinalli G, Spennato P, Sainte-Rose C, et al. Chiari malformation in craniosynostosis. *Childs Nerv Syst* 2005;21:889–901
38. Ventureyra EC, Aziz HA, Vassilyadi M. The role of cine flow MRI in children with Chiari I malformation. *Childs Nerv Syst* 2003;19:109–113

Rehabilitation of Patient of Glass Cut Injury With Forehead Flap Repositioning and Implant-Retained Nasal Prosthesis

Gunjan Pruthi, BDS, MDS, Kirti Bansal, BDS, MDS,* Rakesh Sharma, MBBS, MS,† Ruchi Goel, BDS, MDS,* Nidhi Mangtani, BDS, MDS,‡ and Veena Jain, BDS, MDS‡*

Abstract: Traumatic injuries, especially in maxillofacial region, not only lead to physical debilitation but also cause severe psychological distress in the affected individuals. Complete cosmetic and functional rehabilitation of such patients is a challenging task and thus requires a strategic treatment planning and a multidisciplinary team to execute the treatment. This patient report presents a patient who suffered with a severe glass cut injury leading to massive avulsion of face involving forehead, nose, upper lip, and anterior teeth. Patient was rehabilitated with a combined surgical and prosthetic approach, which involved flap repositioning in forehead, nasal and lip regions and an implant-supported nasal prosthesis to replace missing nose. Missing anterior teeth were replaced with fixed dental prostheses.

Key Words: Facial trauma, flap, implants, maxillofacial prosthesis

From the *Centre for Dental Education and Research (CDER), All India Institute of Medical Sciences (AIIMS); †All India Institute of Medical Sciences (AIIMS); and ‡Department of Prosthodontics, Centre for Dental Education and Research (CDER), All India Institute of Medical Sciences (AIIMS), New Delhi, India.
Received September 15, 2015; final revision received March 3, 2016.
Accepted for publication March 9, 2016.
Address correspondence and reprint requests to Dr Veena Jain, BDS, MDS, Department of Prosthodontics, CDER, AIIMS, New Delhi 29, India; E-mail: jainveena1@gmail.com
The authors report no conflicts of interest.

ISSN: 1049-2275
DOI: 10.1097/SCS.0000000000002733

Midfacial defects may be due to congenital defects, trauma, tumor, or infection. The rehabilitation of midfacial defects always poses a challenge and needs a team approach that includes surgeon, anaplastologist, speech therapist, and maxillofacial prosthodontist.[1]

Among midfacial defects, nasal defects produce significant cosmetic concern as it is the most prominent feature on the face.[2] When the reconstruction of nasal defects with surgical grafts is not possible, the prosthetic reconstruction is used to rehabilitate such defects.[3] The cosmetic success of nasal prosthesis depends upon harmony, texture, color matching, blending of tissue interface, and most importantly the retention of the prosthesis.[4]

Various means of retention have been described for nasal prosthesis like strings or straps anchored behind the head,[5], anatomic undercuts,[6] spectacle frames,[6,7] adhesives,[8] magnets,[9,10] and osseointegrated implants.[11,12] Each one has inherent functional or esthetic limitations. The best method that provides satisfactory retention without compromising form and function is the osseointegrated implants. The implant-retained prosthesis restores the tissue defect for maximum improvement in long-term function, comfort, and esthetics.[1,13,14]

Previous literature has patient reports mostly of patients with partial rhinectomy defects that occurred due to resection of neoplasms.[6,7,9–12] This clinical report presents a challenging patient with large mid-facial defect involving forehead, nose, upper lip, and avulsed/fractured anterior teeth because of glass cut injury. Autogeneous grafting was carried out to rehabilitate upper lip and forehead followed by an implant-retained silicone nasal prosthesis. All the important steps including impression making, fabrication of interim prosthesis, implant placement, clinical, as well as laboratory procedures involved in fabrication of definitive prosthesis have been discussed in detail.

PATIENT DESCRIPTION

A 25-year-old male patient suffered from mid facial trauma when a thick sheet of glass fell vertically from overhead leading to massive bilateral facial avulsion including nose and upper lip. Eventually, the patient suffered with fracture of nasal bone and fracture/avulsion of maxillary and mandibular anterior teeth. The emergency management was done at a nearby hospital where the forehead flap was used to cover the nasal defect. Needle hubs had been inserted in the nostril region to help the patient in breathing (Fig. 1A). Patient's life was saved but this led to significant deformity of face. Patient was then referred to our department for complete esthetic rehabilitation of face, especially nose. Patient was psychologically very distressed because of his unpleasing appearance. Associated chief complaints were difficulty in breathing and missing upper and lower front teeth.

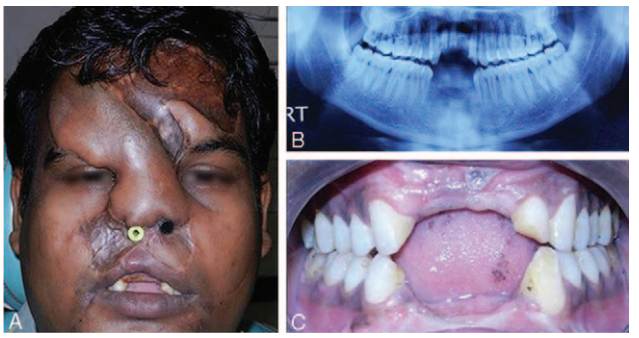

FIGURE 1. (A) Pretreatment extra oral view. (B) Pretreatment ortho pantomogram. (C) Pretreatment intraoral view.