Clinical Significance of Venous Anomalies in Syndromic Craniosynostosis

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Background: The pattern of cranial venous drainage in syndromic craniosynostosis is unpredictable and not adequately understood. Collateral channels substitute for stenotic venous sinuses and pose potential risk for surgical intervention. The purpose of this study was to analyze the patterns of venous drainage in patients with syndromic craniosynostosis and their influence on operative planning and morbidity.

Methods: A retrospective study of patients with syndromic craniosynostosis from 2000 to 2013 was performed. Demographic data were collected including phenotype and associated pathologies. Pre- and/or postoperative venous imaging was reviewed for venous sinus stenosis, collateral emissaries, and persistent fetal sinuses. Categorization of anomalous venous drainage was performed, and the relationship with surgical morbidity was assessed.

Results: Forty-one patients were identified. Anomalies were present in 31 patients (76%) consisting of dural sinus stenosis in 28 (68%), dilated emissaries in 26 (63%), and fetal sinuses in 7 (17%). Pfeiffer syndrome was most commonly associated with anomalous drainage (100%). Venous anomalies were associated with elevated intracranial pressure (ICP), shunted hydrocephalus, Chiari malformations, and sleep apnea. In 5 cases, the surgical plan was adjusted based on anomalous anatomy. No mortalities occurred. Intraoperative complication rate was 7.3%, all with anomalous drainage. Median estimated blood loss was 1,100 cc for patients with anomalies versus 400 cc without anomalies ($P = 0.181$).

Conclusion: Cranial venous anomalies are commonly detected in patients with syndromic craniosynostosis and may affect surgical morbidity and outcome with a higher estimated blood loss, alteration of procedure, and postoperative morbidity. Detailed preoperative imaging of the venous drainage is therefore recommended in cases of syndromic synostosis. *(Plast Reconstr Surg Glob Open 2018;6:e1613; doi: 10.1097/GOX.0000000000001613; Published online 18 January 2018.)*

INTRODUCTION

In the world of craniofacial surgery, patients with syndromic craniosynostosis (SCS) represent some of the most complex and challenging conditions to manage. Multiple suture synostosis, orbital and midface anomalies, anatomic heterogeneity, and functional concerns (airway, intracranial hypertension, feeding, visual and neurodevelopmental) commonly coexist in this population.

Anomalous intracranial venous anatomy is a well-described phenomenon in SCS with potential significance with respect to surgical morbidity.[1–3] It has been proposed that dural venous sinus (DVS) stenosis is a result of skull base abnormalities (ie, jugular foramen stenosis), which leads to compensatory development of collateral emissary veins and persistence of embryologic sinuses.[4] These dilated tributaries may serve as the primary source of cerebral venous drainage.[5] Disruption of these variant vessels during surgery may result in significant, sometimes fatal, intraoperative hemorrhage or intractable venous hypertension secondary to sacrifice of anomalous veins.[2,3]

The literature is sparse when detailing the specific patterns of intracranial venous anomalies (IVA) in SCS; it consists of 2 small studies that describe the patterns

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of IVA in this population[6–8] and a case report of fatal outcome following transection of large emissary veins.[3] Some reports have described cases in which the operative plan was changed based on preoperative vascular imaging.[1,2]

The purpose of this study was 2-fold: to document the patterns of venous anomalies in patients with syndromic craniosynostosis and to analyze their influence on operative planning and morbidity.

METHODS

Following Research Ethics Board (REB) approval (#1000046113), a retrospective review was conducted of all patients with SCS seen at the Hospital for Sick Children, Toronto, from January 2000 to January 2013. Sixty patients were identified for review. Medical records were reviewed for demographic information, phenotypical diagnosis, associated diagnoses, number and type of cranial vault procedures, surgical details, and clinical outcome. Venous anatomy was analyzed directly and through assessment of neuroradiology reports from pre- and/or postoperative computed tomography venography (CTV), magnetic resonance imaging (MRI), and/or magnetic resonance venography (MRV).

Anomalous venous anatomy was assessed by DVS and jugular foramen stenosis, presence of transosseous emissary veins and collaterals, and persistence of fetal sinuses. Preoperative and intraoperative change of surgical procedure, estimated blood loss (EBL), and intraoperative and postoperative complications were also recorded. Information was obtained from imaging studies, radiological and operative reports, and clinical notes.

Definitions of IVA are provided in Figure 1[9] and Figure 2. A scale for DVS stenosis was created (Table 1) to allow for clinical categorization and to aid in analysis
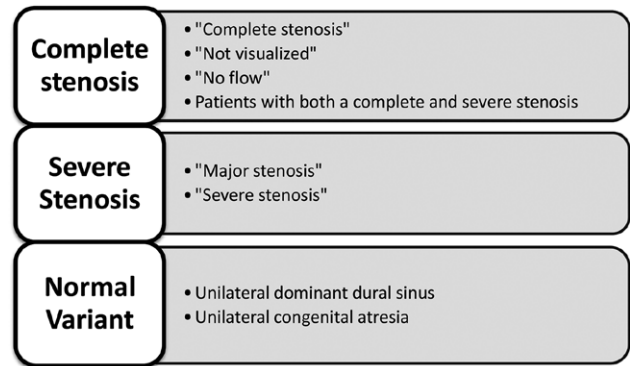

Fig. 1. Definitions of venous anatomy.[9]

Fig. 2. Definition of DVS stenosis based on radiographic findings.

Table 1. Classification of Degree of Dural Sinus Stenosis into 3 Types Based on the Cranial Proximity of the Most Severe Stenosis

Type	Radiographic Finding
I	Severe transverse sinus stenosis (unilateral or bilateral)
	Severe sigmoid sinus stenosis (unilateral)
II	Transverse or sigmoid sinus complete stenosis (unilateral or bilateral)
	Severe sigmoid sinus stenosis (bilateral)
	Severe jugular foramen stenosis (unilateral)
III	Severe jugular foramen stenosis (bilateral)
	Jugular foramen complete stenosis (unilateral or bilateral)

of associated risk factors and predicted clinical outcome (Fig. 3). Data analysis was performed using SPSS software (IBM Corp, SPSS Statistics for Windows, Version 20.0. Armonk, N.Y.). Frequencies and descriptive statistics were examined while comparisons between means and medians were performed with parametric or nonparametric tests accordingly. Chi-square analogs (Cramer V test) were applied for crosstab analysis between multiple nominal and ordinal variables.

RESULTS

Sixty SCS patient files were reviewed. Nineteen files had incomplete data and were excluded. The remaining 41 patients (19 male and 22 female), with a median age at the time of venography of 2 years (mean = 5 years; range = 0.08–17 years) had syndromic pathology in the following frequencies: Apert (11), Crouzon (19), Pfeiffer (7), Saethre-Chotzen (3), and Muenke (1). Patients had the following associated diagnoses: elevated intracranial pressure (37%), ventriculomegaly (22%), shunted hydrocephalus (37%), Chiari malformation (51%), syringomyelia (17%), and obstructive sleep apnea (49%). At the time of review, 38 patients (93%) had undergone at least 1 and 26 (63%) had undergone at least 2 cranial vault procedures (median = 2; range = 0–5). These included 7 suboccipital, 7 posterior vault, 36 anterior vault, and 10 combined anterior and posterior vault reconstructions. Twenty-one patients had CTV, 25 had MRV, and 38 had MRI. There were no complications related to sedation for MRI. No mortalities were encountered.
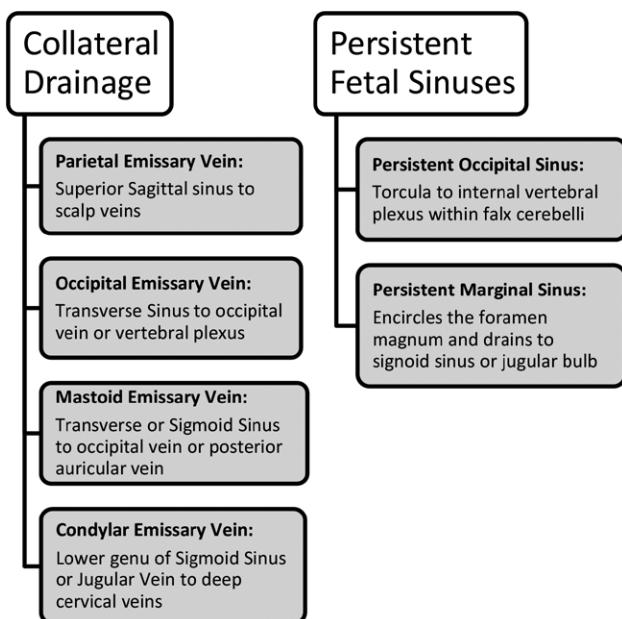
Description of IVA by Syndrome

Thirty-one patients (76%) were noted to have IVA in the form of DVS stenosis, jugular foramen stenosis, collateral emissary drainage, or persistent fetal sinuses (Fig. 4). Documented IVA were present in 7 patients with Pfeiffer syndrome (100%), 16 patients with Crouzon syndrome (84%), and less commonly in Apert syndrome (64%) and Saethre-Chotzen (33%) patients. None were detected in the single patient with Muenke syndrome (Table 2). The number of anomalies by syndrome is demonstrated in Table 3. Representative images of anomalies in a patient with Crouzon syndrome are demonstrated in Figure 5.
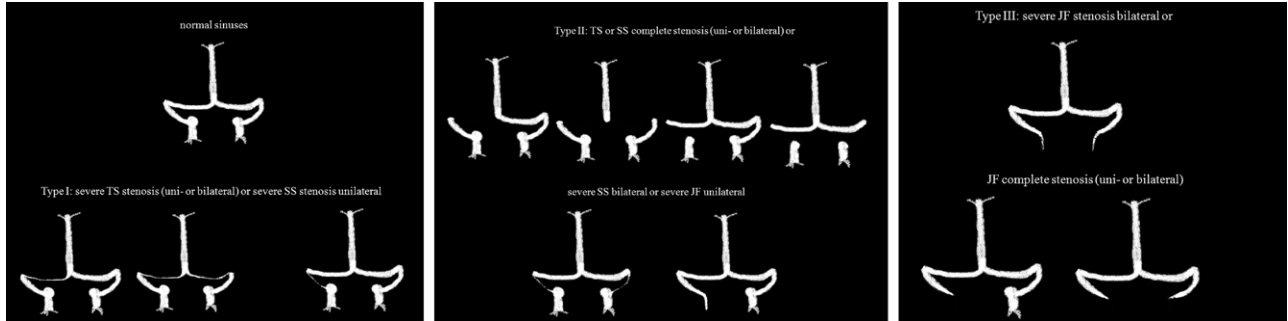
Fig. 3. Representative imaging of DVS stenosis grouped according to type based on proximity to the jugular bulb.
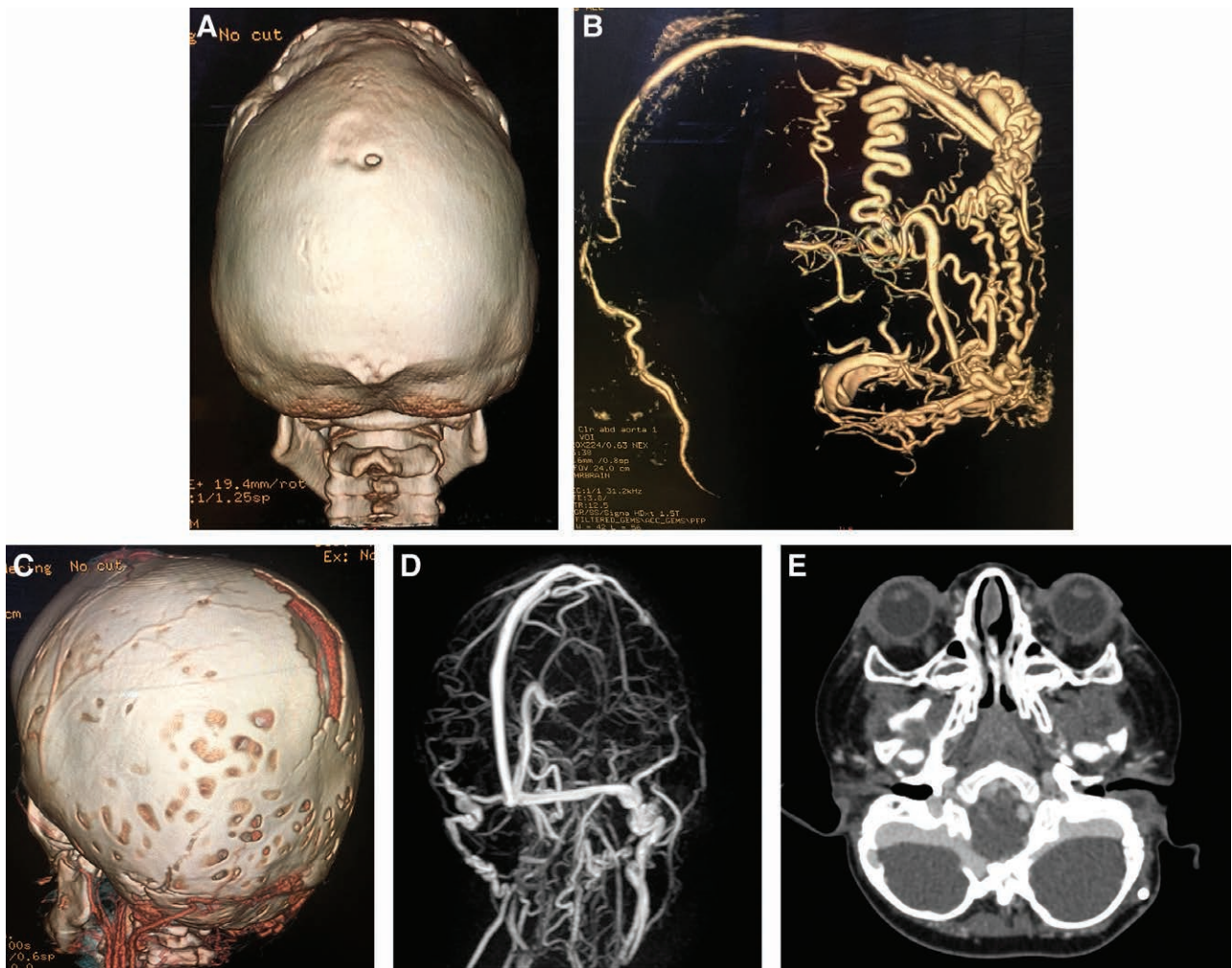
Fig. 4. Representative images of venous anomalies. A, CT scan of an 8-year old male with Crouzon syndrome demonstrating a small innocuous foramina in the midline. B, CT venogram of the same patient showing extensive subcutaneous venous plexus draining from the sagittal sinus in the same patient. C, Emissary veins in the occipital region in a 9-month old female with Pfeiffer syndrome. D, MR venogram showing retrosigmoid emissary veins and jugular foramen stenosis. E, Axial CT venogram demonstrating occipital sinus and jugular stenosis.

Table 2. Venous Anomaly by Syndrome

Diagnosis	Apert	Crouzon	Pfeiffer	SC	Muenke	Total
Total no. patients	11	19	7	3	1	41
Anomalous venous anatomy (%)	7 (64)	16 (84)	7 (100)	1(33)	0 (0)	31 (76)

SC = Saethre-Chotzen.

Dural Venous Sinus Stenosis

Twenty-eight patients (68%) had stenosis of the DVS. Stenosis was bilateral in 22 and unilateral in 6 patients. Stenosis occurred in the transverse sinus (n = 2), the sigmoid-jugular complex (n = 5), or both (n = 21). Stenosis was severe in 5 and complete in 7 patients. Twenty patients (49%) had concomitant jugular foramen stenosis, which was bilateral in 15 and unilateral in 5 patients (Table 4). The frequency of DVS stenosis was 100% for Pfeiffer, 74% for Crouzon, 55% for Apert, 33% for Saethre-Chotzen, and 0% for Muenke syndrome patients. The frequency of jugular foramen stenosis was 57% for Pfeiffer, 55% for Apert, 47% for Crouzon, 33% for Saethre-Chotzen, and 0% for Muenke syndromes (Table 5).

Based on the grading system described in the Methods section, DVS stenoses were 18.2% type I, 27.3% type II, and 54.5% type III. DVS type was assessed in relation to syndrome (Fig. 6). Using the Cramer's V measure, there was no significant association between the type of DVS stenosis and syndrome. The value was 0.395 (0 represents no association and 1 represents strong association), but this finding was not statistically significant ($p = 0.244$).

Collateral Venous Drainage

Dilated emissary veins were present in 26 patients (63%) with the following frequencies: 18 occipital (44%), 16 mastoid (39%), 3 condylar (7%), and 3 parietal (7%; Table 4). With respect to association between transosseous drainage and syndrome, emissaries were found in 100% of Pfeiffer's, 74% of Crouzon's, 36% of Apert's, 33% of Saethre-Chotzen, and 0% of Muenke's patients. Occipital and mastoid emissaries were most common. Parietal emissaries were found predominantly in Crouzon's patients (2/3; Table 4).

An ordinal representative variable was created for the number of emissaries per patient (none, 1–2, and ≥ 3). The percentage of patients per category was 36.6% for none, 48.8% for 1–2, and 14.6% for more than 3 emissaries (Fig. 7). The Cramer's V measure was 0.392, indicating a weak association, without statistical significance ($P = 0.127$).

The relationship between DVS type and number of emissaries was analyzed and showed that all cases with more than 3 emissaries (n = 5) had type III (jugular foramina type) DVS. In addition, 50% of the cases that had 1–2 emissaries again came under the type III category of DVS stenosis (Fig. 8). Although there was an evident trend with respect to increased emissary formation with more distal DVS stenosis, statistical analysis did not demonstrate a strong association (Cramer's V measure = 0.293) or significance ($P = 0.224$).

Persistent Fetal Sinuses

A persistent occipital sinus was found in 7 patients (17%) (5 Crouzon and 2 Pfeiffer) and a persistent marginal sinus in 1 patient (Pfeiffer; Table 4). No persistent fetal sinus was found in any patient with Apert, Saethre-Chotzen, or Muenke syndrome (Table 5).

Impact of IVA on Surgical Planning

The surgical plan was changed preoperatively in 3 patients and intraoperatively in 2 patients based on abnormal venous anatomy (12% of cases) and was limited to patients with Pfeiffer and Crouzon syndromes. Adjustments included limiting the extent of the craniotomy to avoid collateral drainage, limiting openings over persistent sinuses, and preserving significant emissaries (Table 6).

Impact of IVA on Clinical Outcome

Patients with IVA were more likely to have elevated ICP ($P = 0.0022$), ventriculomegaly or shunted hydrocephalus ($P = 0.0033$), Chiari I malformations ($P = 0.0232$), and obstructive sleep apnea ($P = 0.0234$). The association between IVA and the presence of a syrinx was not significant ($P = 0.1551$). We also analyzed the type of anomalies seen in patients with versus without elevated ICP (Table 7). The overall intraoperative complication rate was 7.3% (3/41). Complications included 1 dural tear resulting in persistent cerebrospinal fluid (CSF) leak and infection, 1 case with significant bleeding, and 1 that presented with intraoperative brain swelling. These complications occurred in 3 of 31 patients with IVA compared with 0 of 10 patients without anomalies; however, this finding was not significant ($P = 0.3069$). There were no postoperative complications and no mortalities.

We also examined the EBL as it was recorded in the anesthetic record. We calculated the total EBL as a sum of all procedures for each patient. Data were available for 25 patients. Median EBL in patients with IVA was 1,100 cc, whereas in patients without anomalous drainage, it was 400 cc. Although this implies a trend, the difference was not significant ($P = 0.181$). We further examined the potential significance of anomalous drainage patterns by analyzing

Table 3. Number of Venous Anomalies by Syndrome

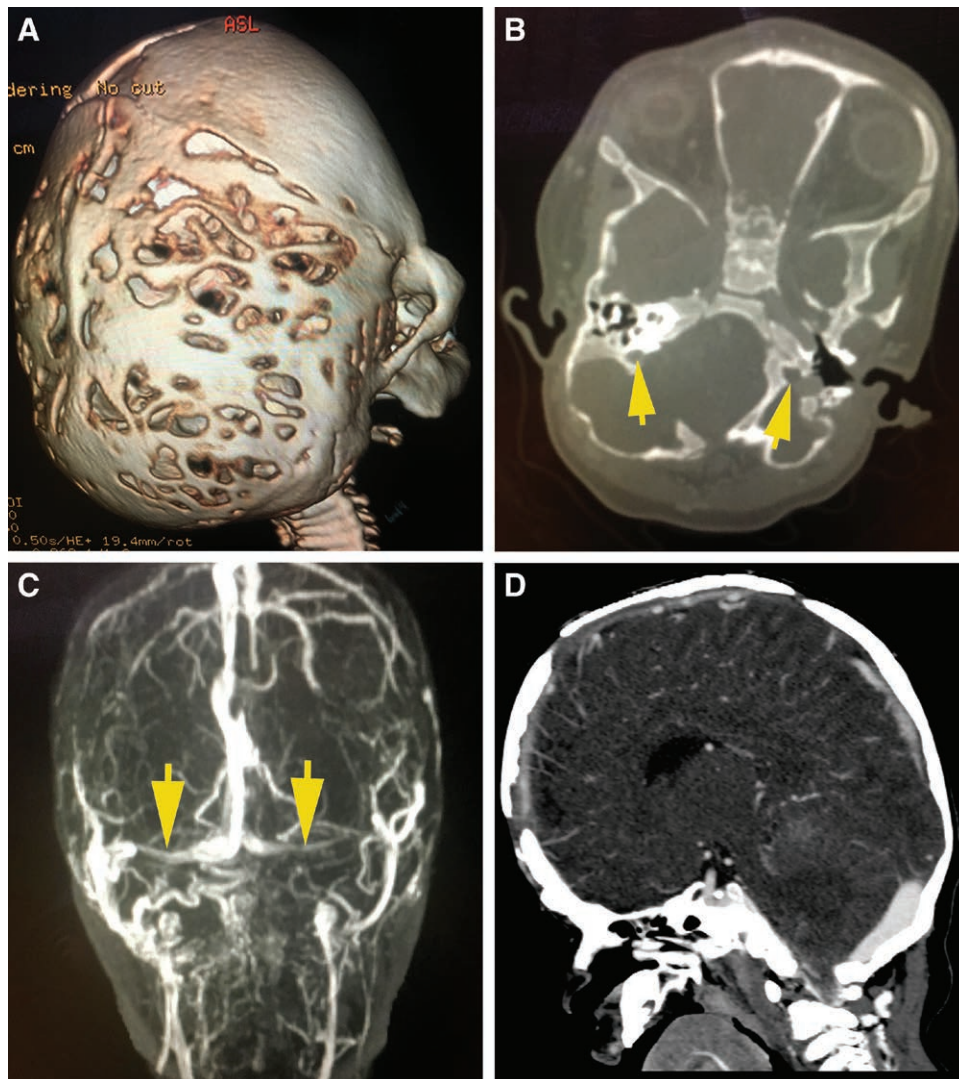| Syndrome	No. Anomalies									Mean
	0	1	2	3	4	5	6	7	8	
Apert	1	2	4	3	1	0	0	0	0	2
Crouzon	2	1	2	4	4	3	3	0	0	3
Pfeiffer	0	0	2	1	2	1	0	0	1	4
Saethre-Chotzen	1	1	0	0	1	0	0	0	0	2
Muenke	0	1	0	0	0	0	0	0	0	1

Fig. 5. A 9-month-old female with Crouzon syndrome showing extensive thumbprinting (A), bilateral jugular foramina stenosis (yellow arrows) (B), hypoplasia of the transverse and sigmoid sinuses, bilaterally, mastoid emissary vein and collaterals (yellow arrows) (C), and persistent occipital fetal sinus (D).

Table 4. Frequency of Venous Anomalies

Venous Anomaly	No. Patients (%)
DVS stenosis	28 (68)
Bilateral	22 (54)
Unilateral	6 (15)
TS only	2 (49)
SJC only	5 (12)
TS and SJC	21 (51)
Severe stenosis	5 (12)
Complete stenosis	7 (17)
Jugular foramen stenosis	20 (49)
Bilateral	15 (37)
Unilateral	5 (12)
Transosseous emissary veins	26 (63)
Mastoid	18 (44)
Occipital	16 (39)
Condylar	3 (7)
Parietal	3 (7)
Persistent fetal sinuses	8 (20)
Occipital	7 (17)
Marginal	1 (2)

TS = transverse sinus; SJC = sigmoid-jugular complex.

the association between EBL and DVS stenosis, number of emissaries, and presence of persistent fetal sinuses. The median EBL was 1,500 cc for type I, 200 cc for type II, and 1,200 cc for type III stenosis ($P = 0.180$). Similarly, the median EBL was 600 cc when there were no emissary veins present, 1,450 cc when there were 1–2, and 900 cc for more than 3 emissaries ($P = 0.568$). Regarding the presence of fetal sinuses, we compared median EBL with persistent sinuses via the Mann-Whitney U test and did not find a statistically significant difference, although there was a clear trend with 1,250 cc in cases with persistent fetal drainage versus 800 cc in cases without ($P = 0.184$; Table 8). Mean EBL was compared with procedure type/location with no significant association ($P = 0.214$; Fig. 9).

DISCUSSION

To our knowledge, this is the largest documented cohort of patients with syndromic craniosynostosis. Anoma-

Table 5. Frequency of Venous Anomalies by Syndrome

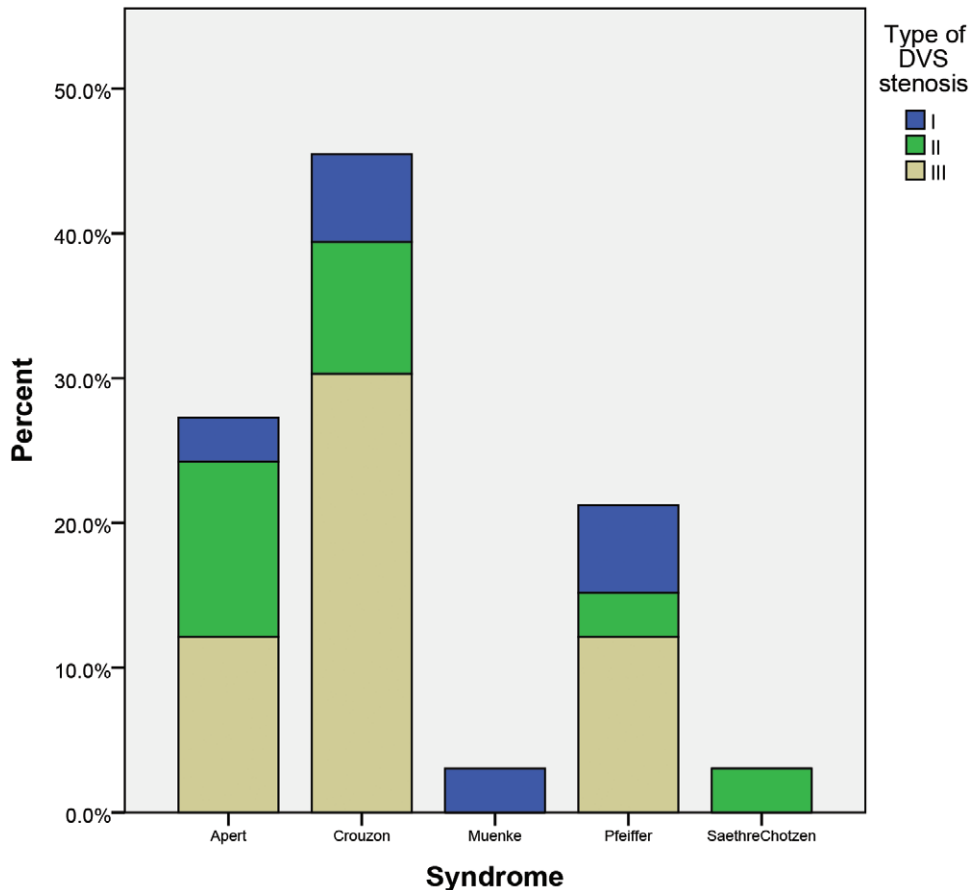| Diagnosis n (%)	Pfeiffer (n = 7)	Crouzon (n = 19)	Apert (n = 11)	SC (n = 3)	Muenke (n = 1)
Venous anomalies	7 (100)	16 (84)	7 (64)	1 (33)	1 (100)
DVS stenosis	7 (100)	14 (74)	6 (55)	1 (33)	0
JF stenosis	4 (57)	9 (47)	6 (55)	1 (33)	0
Emissary veins	7 (100)	14 (74)	4 (36)	1 (33)	0
Persistent fetal sinuses	3 (43)	5 (26)	0	0	0

JF = jugular foramen.

Fig. 6. DVS stenosis type according to syndrome.

lous intracranial venous anatomy is well described in this population.[5] The increased recognition of these anomalies carries significance with respect to surgical planning and morbidity. Venous hypertension due to outflow obstruction increases the risk of significant blood loss during reconstructive surgery and contributes to the development of hydrocephalus with a subsequent increased risk for postoperative CSF leak.[3,10] In the setting of severe DVS stenosis, transosseous collaterals and persistent fetal sinuses may represent the primary route of venous drainage. These anomalous vessels pose significant surgical risks, including intraoperative hemorrhage, air embolism, and potentially lethal venous hypertension due to inadvertent closure of transosseous collaterals.[3,11] Although there are risks to imaging in the form of ionizing radiation (CT) and general anesthesia (MRI), these risks are warranted, given the potential for significant morbid-

ity and mortality if this information is not included in the surgical planning.

The results of our study and of others demonstrate that preoperative planning must include an evaluation of IVA in most cases. Thompson et al.[3] provided a salient example of the potential for significant surgical morbidity in an 8-year-old girl with Pfeiffer syndrome who suffered fatal intracranial hypertension after a large occipital emissary was sacrificed during the scalp incision for a cranial vault expansion. At autopsy, it was discovered that there were severe, bilateral stenoses of the jugular foramina and the occluded occipital emissary vein was the primary route of drainage. There are several other reports in which the detection of IVA on preoperative vascular imaging led to the cancelation of posterior vault expansion procedures.[1,2,6] No large studies currently exist, however, providing a description of the types of anomalies seen by syndrome, and
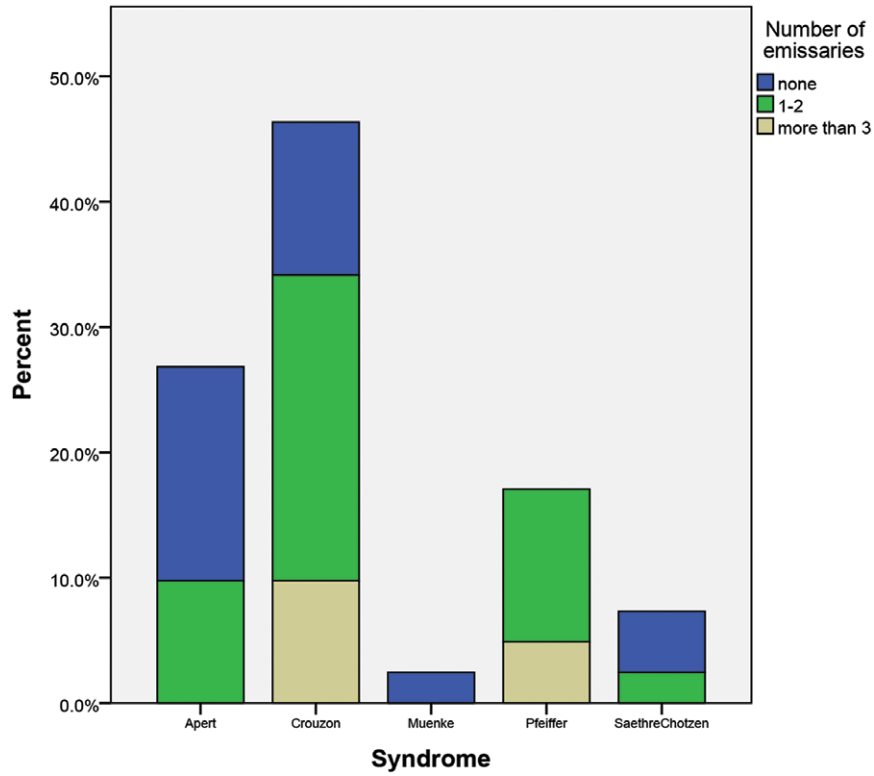
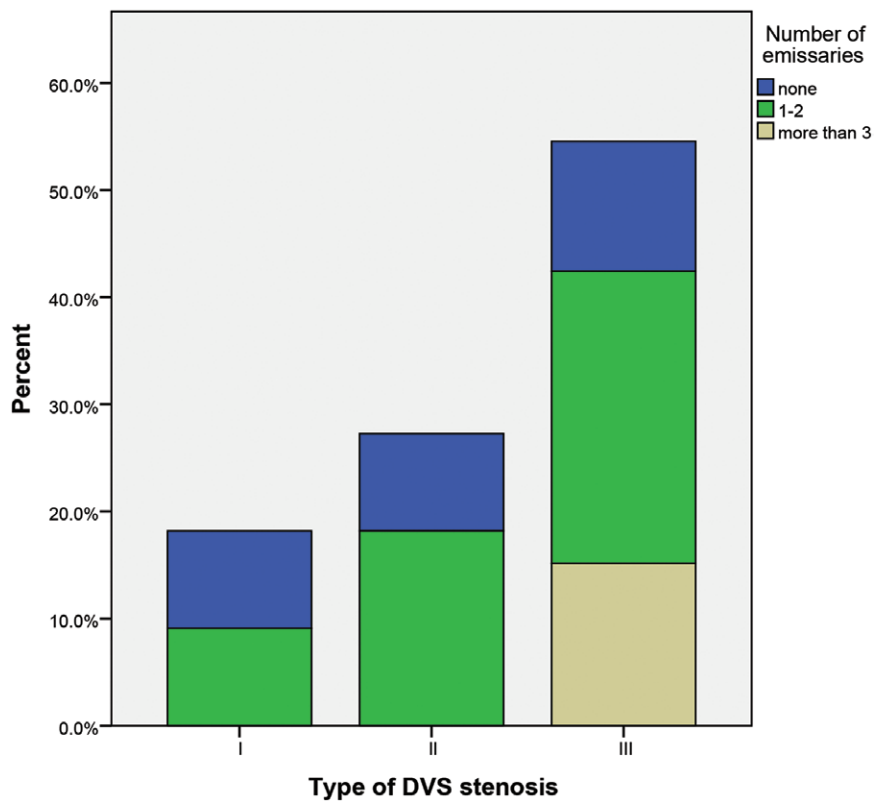**Fig. 7.** Percentage of collateral emissaries by syndrome.

Fig. 8. Number of venous emissaries and association with proximity of dural sinus stenosis, or DVS type.

Table 6. Summary of Cases in Which Operative Plan Was Changed Either Pre- or Intraoperatively

Diagnosis	Age (mo)	Procedure	Anomalies	Associated Conditions	Change in Plan	Complications
Pfeiffer	2	Total vault reconstruction	Bilateral stenosis of the sigmoid sinuses, jugular complexes, and jugular foramina and absent left transverse sinus. Large bilateral occipital emissaries, persistent occipital sinus, and a right transverse sinus draining to superficial scalp veins.	Elevated ICP, shunted hydro-cephalus, Chiari I malformation with syrinx, and obstructive sleep apnea.	(Preoperative); Procedure was altered to avoid the occipital region.	None
Pfeiffer	7	Posterior vault distraction	Bilateral occipital emissaries. Bilateral stenosis of the jugular bulbs, hypoplastic left transverse and sigmoid sinuses, and persistent occipital sinuses.	Elevated ICP, shunted hydrocephalus, and Chiari I malformation.	(Preoperative); Procedure was altered to preserve occipital emissaries.	CSF leak and epidural infection.
Crouzon	24	Posterior vault distraction	Transosseous emissaries near the torcula with bilateral stenosis of transverse and sigmoid sinuses and jugular complexes.	Elevated ICP, ventric-ulo-megaly, Chiari I malformation, and obstructive sleep apnea	(Preoperative); Distraction remained above the torcula and emissaries were preserved.	None
Crouzon	145	Monobloc oste-otomy	Parietal and occipital emissaries.	Chiari I malformation and obstructive sleep apnea.	(Intraoperative); Procedure altered to preserve scalp collaterals	None
Pfeiffer	29	Posterior cranial vault distraction	Right transverse, right sigmoid, and right jugular complex stenosis. Narrow left sigmoid and left jugular complex. Occipital emissaries.	Elevated ICP and ventriculo-megaly	(Intraoperative); Distraction was limited to the superior nuchal line due to emissaries near the torcula.	Skull fracture and small epidural hematoma diagnosed postoperatively. No morbidity related to venous hypertension or intraoperative hemorrhage.

Table 7. Types of Venous Anomalies Seen in Patients with Versus without Elevated ICP

Venous Anomaly n (%)	With Elevated ICP (n = 15)	Without Elevated ICP (n = 26)	P
DVS stenosis	14 (93)	20 (77)	0.2324
Emissary veins	14 (93)	12 (46)	0.0027
Fetal sinuses	3 (2)	3 (12)	0.6509

Table 8. Association between EBL and Venous Anomalies

Venous Anomaly	EBL, Median and IQR (cc)	P
With anomalies (n = 24)*	1,100 (1,443.8)	
Without anomalies (n = 5)*	400 (882.5)	0.181
No emissaries (n = 8)†	600 (1,050)	0.568
1–2 Emissaries (n = 17)†	1,450 (1,575)	
> 3 Emissaries (n = 4)†	900 (687.5)	
Type I (n = 3)‡	1,500	0.180
Type II (n = 7)‡	200 (1,117)	
Type III (n = 15)‡	1,200 (1,650)	
(+) Fetal sinuses (n = 8)§	1,250 (818.8)	0.184
(−) Fetal sinuses (n = 21)§	800 (1,300)	

*Total EBL in patients with and without anomalous venous drainage (Mann–Whitney U test).
†Total EBL in patients according to the number of emissaries (Kruskall–Wallis test).
‡Total EBL in patients according to the type of DVS stenosis (Kruskall–Wallis test).
§Total EBL in patients with and without persistent fetal sinuses (Mann–Whitney U test).
EBL, estimated blood loss; IQR, interquartile range.

the relationship to outcome with respect to surgical planning and potential morbidity. In children, the radiation or general anesthesia required to obtain imaging necessitates that any study be justified. As a result, we assessed the incidence of IVA by syndrome and the clinical significance of these anomalies.

In our study, anomalous IVA was identified in 76% of patients. Both collateral drainage and DVS stenosis were common. Abnormal transosseous veins were observed in 63% of patients. The most common anomalous collaterals were occipital and mastoid emissary veins, posing significant surgical risk for posterior vault and suboccipital procedures. Condylar and parietal emissaries were less common; however, knowledge of their frequency and most associated syndromes is important as they are relevant for more anterior or combined approaches. In addition to these transosseous anomalies, we noted significant abnormalities in the normal intracranial drainage mechanisms.

Twenty-eight patients (68%) had DVS stenosis, which was most commonly bilateral (78%) and involving the sigmoid-jugular complex (98%). Twenty patients (49%) had jugular foramen stenosis, which was severe or complete in all patients, bilateral in the majority (75%) and was always associated with DVS stenosis. Persistence of occipital and marginal fetal sinuses was less frequent, only occurring in 19% of patients.
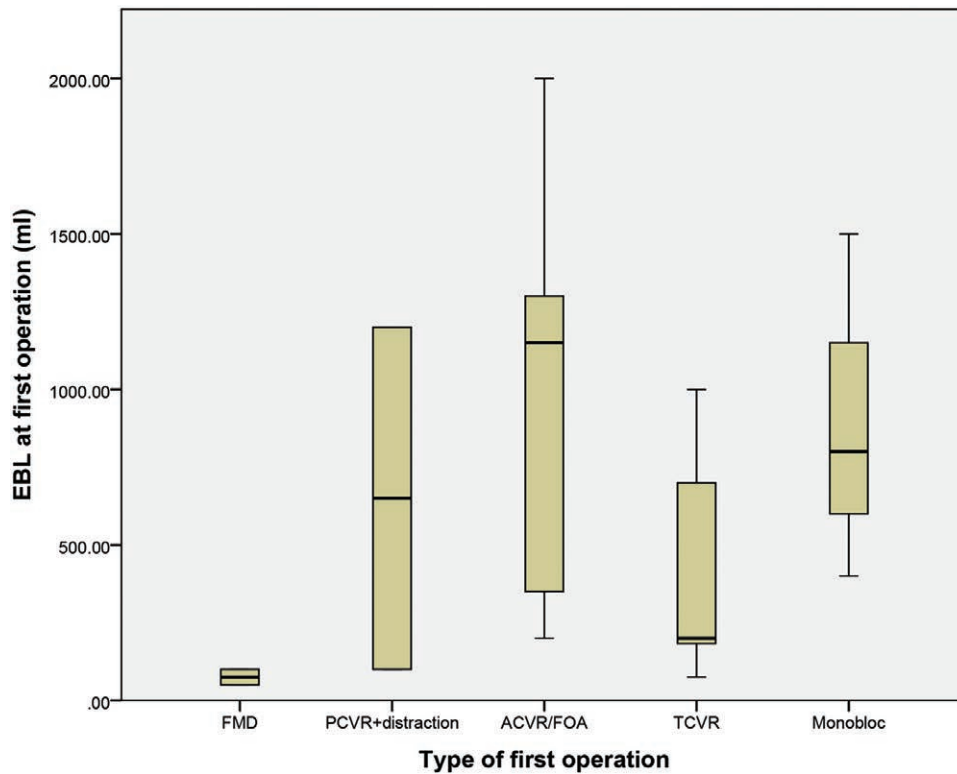
Fig. 9. Association between EBL and procedure type. Using the Kruskall–Wallis test to compare means for each type of procedure, there was no significant difference in EBL by surgery ($P = 0.214$). FMD + foramen magnum decompression; PCVR = posterior cranial vault reshaping; ACVR = anterior cranial vault reshaping; FOA = fronto-orbital advancement; and TCVR = total cranial vault reshaping.

Frequency of IVA by syndrome is important in informing preoperative work-up and assessment. We found that IVA, were most common in Pfeiffer (100%) and Crouzon (84%) and slightly less common in Apert patients (64%). We cannot make any statements as to associations with Saethre-Chotzen and Muenke syndromes as there were only 3 patients and 1 patient in our cohort, respectively. Given these numbers overall, we recommend preoperative vascular imaging in all syndromic patients.

Many reports have demonstrated similar variability in venous drainage in children with complex synostosis. Jeevan et al.[6] demonstrated that of 11 patients with syndromic synostosis, 9 had enlarged emissary veins, and 4 of these patients had a transosseous route as their primary route of drainage on CTV. Taylor et al.[8] studied 23 synostosis patients, 18 of whom were syndromic and found that 17 patients had either severe stenosis (51–99%) or no flow in the sigmoid sinus and/or jugular complex by digital subtraction angiography. In 11 patients, significant venous collaterals were seen in the region of the stylomastoid emissary vein. In a study done by Rollins et al.[7], MRV showed jugular stenosis and transosseous venous drainage in 12/17 patients with complex craniosynostosis, only 11 of which were syndromic. The most common emissary veins were posterior condylar, seen in 11/17 patients. These results contrast with the present study where occipital (44%) and mastoid (39%) emissaries were seen most frequently, with condylar emissaries being observed

in only 7% of patients. These condylar emissaries were most common in Crouzon syndrome (67%). This may be due to differences in patient population studied as 6/17 patients were nonsyndromic in the Rollins et al.[7] paper.

With respect to the significance of IVA in the present study, morbidity was not significantly associated with IVA. Although there was a trend toward increased EBL based on the number of emissaries and DVS type, these findings were not significant. In addition, although there was a significant relationship between IVA and elevated ICP, shunted hydrocephalus, Chiari malformations, and obstructive sleep apnea, there was no significant difference in complication rate between patients with and without IVA. Surgical morbidity was more closely related to the associated conditions than to the anomalous drainage itself. The skull fracture and resulting epidural hematoma, for example, was most likely secondary to significant thinning of the skull from chronic hydrocephalus. Similarly, the CSF leak was likely related to hydrocephalus and elevated ICP.

Although no procedures were abandoned in the present study, there were 5 cases in which the procedure was altered to accommodate the venous anatomy. This is compared to the study by Jeevan et al.[6] in which abnormal venous anatomy led to the abandonment of operative intervention in 4, and a change in the planned procedure in an additional 3 cases. Importantly, however, all our study patients had undergone preoperative venous imaging, therefore allowing for preoperative planning. Compari-

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son of outcomes with and without venous imaging would be more informative with respect to the true morbidity associated with these radiographic findings. However, with the current accrual of information surrounding the prevalence of IVA in syndromic synostosis, a controlled prospective study would be unethical. Additionally, the association between the type and location of procedure and EBL was investigated and found to be insignificant. This further underscores the fact that the trend toward an association between blood loss and IVA may be independently significant in a larger cohort as it cannot be explained simply by the type of surgery performed.

Our study was limited by the fact that we did not have control subjects to assess normal variation in venous drainage. Instead, our definitions of "anomalous" versus "not anomalous" were based on the current literature and neuroradiology reports. Additionally, since this was a retrospective review, the number of previous cranial vault procedures and VP shunts at the time of venography was not standardized. Therefore, whether the venous anatomy had been affected by prior operative intervention is not known. However, Jeevan et al.[6] studied 11 patients with syndromic craniosynostosis using CTV and "found no relation between … the existence of previous cranial surgery and the presence of collateral venous drainage channels." Although cranial vault remodeling does not address jugular foraminal stenosis, it decreases craniocephalic disproportion and intracranial pressure. In this respect, prior intervention may be expected to decrease venous anomalies in the form of emissaries and dural stenoses. However, it is also possible, depending on preoperative anatomy, that sacrificing emissaries intraoperatively would worsen preexisting anomalies and potentially increase the likelihood of related issues such as elevated intracranial pressure and hydrocephalus. This will be an important relationship to study in a standardized, prospective fashion to help inform our understanding of the etiology of these anomalies in craniofacial syndromes.

CONCLUSIONS

This study represents the largest cohort of children with syndromic craniosynostosis and the results demonstrate a high incidence of IVA in this population. These data therefore allow for a more accurate assessment of the most common venous drainage patterns and their clinical significance. Additionally, the categorization of DVS stenosis based on proximity to the skull base or jugular foramen allows for a more standardized method of documentation.

Increased recognition of these anomalies carries significance with respect to surgical planning and reducing morbidity in the treatment of these patients. Based on the incidence and pattern of venous anomalies found in the present study, we recommend venous imaging as part of preoperative planning in all children with syndromic craniosynostosis. Where previous studies have not been able to stratify their recommendations based on phenotype due to small sample sizes, we assert that preoperative venous imaging is most crucial in patients with Pfeiffer, Crouzon, and Apert syndrome.

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