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# A Two Center Review of Three Techniques for Posterior Vault Expansion Following Either a Staged or Expectant Approach to the treatment of Crouzon and Apert Craniosynostosis

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**Abstract**

**Introduction:**

Timing of posterior cranial expansion for the management of intracranial pressure can be 'staged' by age and dysmorphology or 'expectant' by pressure monitoring. We report shared outcome measures from one center performing posterior vault remodeling (PCVR) or distraction (PVDO) following a 'staged' approach and another performing spring assisted expansion (SAPVE) following an 'expectant' protocol.

**Methods:**

Apert or Crouzon syndrome cases who underwent posterior expansion less than two years old were included. Perioperative outcomes and subsequent cranial surgeries were recorded up to last follow-up and intracranial volume changes measured and adjusted using growth curves.

**Results:**

38 patients were included. Following the 'expectant' protocol, Apert cases underwent SAPVE at a younger age (8 months) than Crouzon cases (16 months). The initial surgery time was shorter but total operative time, including device removal, longer for PVDO (3:52) and SAPVE (4:34) than for PCVR (3:24). Growth-adjusted volume increase was significant and comparable. 14% PCVR, 33% PVDO, and 11% SAPVE cases had complications, but without long-term deficits. Following the staged approach, 5% only underwent PVDO, 85% had a staged posterior followed by anterior surgery, and 10% required a third expansion. Following the expectant approach, 42% patients had only posterior expansion at last follow-up, 32% had a secondary cranial surgery, and 26% had a third cranial expansion.



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**Conclusions:**

Two approaches involving posterior vault expansion in young syndromic patients using three techniques resulted in comparable early volume expansion and complication profiles.

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Children with syndromic craniosynostosis undergo cranial expansion to manage potential, impending or diagnosed increased intracranial pressure (iICP). Incidences of iICP range from 45 - 83%[1,2] in Apert and 61 - 63% in Crouzon syndromes.[3,4] Protocols differ on timing and methods for cranial vault expansion, but some advocate posterior expansion as the initial treatment, allowing frontal surgeries to be performed at a later age.[2,5,6] Timing can be 'staged' based on age,[6,7] or 'expectant' based on close monitoring and treatment once iICP is detected.[2,3] Recognized procedures include one-stage posterior cranial vault remodeling (PCVR),[8,9] posterior vault distraction osteogenesis (PVDO),[10] and spring assisted posterior vault expansion (SAPVE).[11–13] It is rare that a single center will offer all three techniques. In the absence of prospective multi-center trials, retrospective data can instead be collected by multiple centers, recognizing that operative protocols and referral patterns will differ making direct comparison challenging.

Our purpose was to report perioperative variables, early volumetric change, and need for subsequent cranial surgery associated with PCVR and PVDO in a center following a staged approach and with SAPVE at another center following an expectant approach. Our aim was to identify differences and similarities in the anticipated cranial surgeries a patient will undergo in their first decade following these two different approaches.

**METHODS**

Institutional review boards at both hospitals approved this retrospective study. All patients with Apert or Crouzon syndrome were included who underwent either PCVR, PVDO, or SAPVE under the age of two years as a first cranial surgery between 2004 and 2018 and had pre- and post-treatment computed tomography (CT) scans. Patients with prior transcranial procedures before the follow-up scan were excluded.



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**Seattle Children's Hospital (SCH) "Staged" Approach**

Patients diagnosed with syndromic synostosis were assessed for hydrocephalus and signs of iICP with CT in the first three months of age by the interdisciplinary team. Airway evaluation and polysomnography were performed based on symptoms and by a standardized questionnaire.[14] Airway optimization and need for ventriculoperitoneal shunt were prioritized over cranial surgery. Patients with posterior brachycephaly and signs of cephalocranial disproportion on CT underwent prophylactic posterior cranial expansion within the first year.[6] Prior to 2005, all patients were treated with PCVR. Subsequently, one surgeon continued with PCVR, and the other two surgeons switched to PVDO except for patients with severe asymmetry when PCVR would be used. Patients with concomitant anterior brachycephaly underwent fronto-orbital advancement (FOA) around two years of age unless severe exophthalmos necessitated an earlier FOA or an early monobloc distraction procedure.[6] The surgical goal of this staged approach was to maximize intracranial space prophylactically in the first two years of life. Patients were followed with annual visits and fundoscopy. Any symptoms concerning for iICP initiated an escalating protocol that included optical computed tomography (OCT), and if indicated inpatient direct pressure monitoring.[15]

*Posterior Cranial Vault Remodeling (PCVR)*

PCVR was performed through a coronal scalp incision with a sub-galeal dissection from coronal sutures posterior to the skull base. The parietal and occipital bones were dissected from the dura and removed, then orthotopically replaced in an expanded position with resorbable fixation (Figure 1). Blood products were administered from the beginning of the case with the volume determined by estimated blood loss, with a goal not to let the hematocrit drop below 20%. One patient in the study had a post-operative molding helmet placed due to concern over the stability of the expansion from poor quality bone.



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*Posterior Vault Distraction Osteogenesis (PVDO)*

PVDO was performed with the same approach as PCVR. A circumferential osteotomy was performed from the vertex, inferior to the squamosal suture, posterior above the petrous ridge, then inferior to the torcula as low as possible and continuing in a symmetric pattern on the contralateral side. No epidural dissection was performed except for that needed for the osteotomy. Paired commercially available 25-30 mm internal mandible distraction devices were placed parallel to Frankfort Horizontal just superior to the squamosal suture and fixated with 3mm screws. Detachable percutaneous activation arms extend anteriorly (Figure 2). Blood products were administered as per PCVR protocol. One to two days post-operation, activation was begun at a rate of 1-2mm/day until maximum device activation. Although the goal was to fully expand the device, activation was stopped early if there were signs of excessive patient discomfort or skin pressure. The devices were removed through direct parallel incisions under a general anesthesia following a 6-to-8-week consolidation period.

**Great Ormond Street Hospital (GOSH) "Expectant" Approach**

Patients diagnosed with syndromic synostosis underwent CT scan, fundoscopy and polysomnography. Visual evoked potentials (VEPs) were obtained with fundoscopy at ages 3, 4, 6, 9, 12, and 18 months. VEPs are an indirect measure of iICP by detecting the integrity of visual pathway function. VEP latency has been shown to have a strong positive correlation with ICP ($r=0.84$).[16] The team followed an "expectant" approach that treated iICP only after it has been detected by VEP monitoring or suspected on clinical examination.[2,3] If present, hydrocephalus and obstructive sleep apnea were treated first by ventriculoperitoneal shunt or adenoid – tonsillectomy respectively. Subsequent treatment was a first stage SAPVE. Springs were removed during a future planned anesthetic when possible. Subsequent cranial



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expansion was based on VEP monitoring at 3, 4, 6, and 10 years of age, or between these times points if there were any concerning clinical signs or symptoms. iICP was either treated with a repeat posterior expansion or a trans-cranial fronto-facial surgery depending on age and indications.

*Spring-Assisted Posterior Vault Expansion (SAPVE)*

SAPVE was performed through a coronal scalp incision with a subgaleal dissection 7cm posteriorly. A posteriorly based pericranial flap was raised, superior to the temporalis muscles, and two tunnel extensions were dissected towards the petrous ridge, then posterior across the occiput towards the foramen magnum. A curvilinear osteotomy was made starting 5cm posterior to the skin incision and extending along the dissected tunnels. Upon completion, the posterior-based bone flap was dissected from the dura a few centimeters and the 'give' was tested by manual pressure and if indicated the osteotomies were extended further towards the foramen magnum or more dural dissection was performed. Paired custom-made springs[17] were then placed into grooves in the bone approximately two centimeters either side of the midline (Figure 3). Spring strength was chosen by the operating surgeon; if two springs were felt to be insufficient, further springs were placed. The pericranial flap and skin were closed while compressing the springs. The springs were self-activating and estimated to complete maximal opening over a period of ten days. Removal of the springs was performed as a separate procedure under general anesthesia, six to twelve months post-operatively, or combined with another other planned anesthetic.

**Data analysis**

Demographics and peri-operative variables were collected. Subsequent cranial surgeries were recorded up to twelve years of age. Intracranial volume (ICV) was measured on the pre-operative CT closest to the surgery and after device removal for PVDO and



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SAPVE.[18] For PCVR, the post-operative scan closest in timing to the other two groups was chosen. The pre-operative ICV was adjusted using syndrome-specific growth curves to compensate for the variability in time between scans.[19] Complications were graded using the Oxford craniofacial complication scale.[20] Due to small sample sizes, nonparametric tests (Kruskal-Wallis H test for multi-group and Mann-Whitney U test for two-group comparisons) were used to analyze the differences in operative parameters and ICV change by technique and by diagnosis. All statistical analysis was performed in R [21].

**RESULTS**

Thirty-eight patients were included with an average post-expansion follow-up of 8.9 years (range 2.0 – 14.1) for GOSH and 6.9 years (range 2.3 – 15.1) for SCH. This represented 23% of the Apert and Crouzon cases in the SCH database and 35% at GOSH due to the inclusion criteria we established to minimize variability. The majority of cases excluded at SCH were because the first cranial surgery was a frontal cranial advancement due to lack of posterior brachycephaly, and at GOSH the majority were excluded since the first posterior cranial surgery was performed after two years of age due to no earlier signs of iICP (Table 1). Operating times for the two device placement surgeries were comparable (PVDO 2:45; SAPVE 2:38), and shorter than for PCVR (3:24). When the device removal surgery was added, the total mean operative time was significantly longer for PVDO (3:52) and SAPVE (4:34) than for PCVR (Table 2). Following a 1:1 blood replacement protocol, the mean transfusion volumes for PVDO (561.8 ml) and PCVR (646.3 ml) were higher than SAPVE cases (234.9 ml), where replacement was based on intra-operative patient condition. There was no difference between transfusion volume for PVDO and PCVR (Table 2)..

When comparing the operative parameters of the two syndromes (Table 3), SAPVE in Apert patients was performed at a younger age and there was greater transfusion volume than



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in Crouzon cases. Following PVDO, Apert patients stayed longer in hospital (11 days) than Crouzon patients (4 days). The devices were removed sooner following PVDO than SAPVE.

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**Complications**

One in seven PCVR cases (14%), 4/12 PVDO cases (33%) and 2/19 SAPVE cases (11%) had complications (Table 4). The Grade 1 complication post-PCVR was a superficial skin infection which required antibiotic treatment in the one patient with a post-operative molding helmet. Post-PVDO complications were a Grade 1 device-related superficial skin infection resolved with antibiotic, and three Grade 3 complications: one device mechanical failure requiring removal; one CSF leak during device activation which required a dura repair and insertion of a lumbar drain; and one migration of the device activation arm below the skin, requiring an operation to re-expose. There were two Grade 3 complications in the SAPVE cohort: one spring exposure which necessitated early removal; and one retained wound drain which required surgical removal.

**Intracranial Volume (ICV)**

All three techniques achieved significant increases in growth adjusted ICV ($p < 0.05$) with no significant differences between groups overall or by syndrome (Figures 4 and 5, Table 5 and 6).

**Subsequent Cranial Procedures**

Following the staged approach, one Crouzon patient required a repeat PCVR prior to the frontal surgery due to signs of iICP, but none of the PVDO cases had a repeat posterior expansion. All except one Apert patient had staged anterior surgeries: 7 FOAs and 2 monoblocs. All Crouzon patients had staged anterior surgeries: 7 FOAs and 3 monoblocs. One Crouzon patient underwent a monobloc advancement (third cranial surgery) 3 years after their FOA. (Figures 6 and 7) Following the expectant approach, 6/11 Apert and 2/8 Crouzon patients underwent repeat posterior surgery within 2 years after primary SAPVE, three Crouzon patients underwent frontal surgeries around 9 years of age and three Apert patients



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underwent frontal surgeries around 4 years of age for signs of iICP. The remaining patients had not had anterior cranial surgeries at last follow-up.

**DISCUSSION**

We report the outcomes of posterior vault expansion at one tertiary care center following a "staged" approach to syndromic synostosis using PCVR or PVDO as a first cranial procedure and another following an "expectant" approach using SAPVE. Eight of the 19 patients (42%) in SAPVE cohort underwent repeated posterior expansion an average of 17 months after the first surgery based on continued monitoring for iICP, and all but one of the 19 patients following the 'staged' approach had protocolized frontal surgery an average of 11 months after posterior surgery (Figure 6). This data suggests that a second cranial expansion surgery achieved clinical cephalo-cranial proportion in the majority of patients at both centers based on team evaluation at last follow-up (Figure 7).

The complication rates were comparable to those previously reported, with 14% for PCVR, 33% for PVDO and 10% for SAPVE (Table 4). A systematic literature search reported an average complication rate after PVDO of 30% (12.5-100%) with no long-term deficits, similar to our series,.[22] Lauritzen reported a 5% spring dislodgement and 2% repeat surgery rate for his first 100 cases, however among the 7 Crouzon and 5 Pfeiffer patients, 25% had spring dislocation and one experienced skin erosion requiring removal.[13] The estimated blood loss in these syndromic cases was 503 ml compared to 143-228 ml for single suture cases, which is similar to the transfusion volumes recorded in this current study. It is of interest that four of the five Grade 3 (return to the operating room) complications in our joint series were device-related. Although other studies [23] have not observed an increased complication rate between PVDO and PCVR, our study does demonstrate the possibility of device-related complications. This emphasizes the need for continued studies on outcomes



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following PVDO and SAPVE to determine if there are any benefits relative to PCVR that outweigh the increased potential for complications associated with use of a device. To date, other retrospective studies such as ours have yet to demonstrate a consistent benefit of one technique over another.

It is challenging to compare techniques between centers and within a center over time since the surgeries are not done in isolation but as part of protocolized care. Operating times for the two device surgeries (PVDO and SAPVE) were shorter than the PCVR surgery, but when the device removal surgery was added the total mean operative times were longer than PCVR (Table 2), and comparable to previous studies examining these procedures in isolation.[9,13,24,25] We noted no difference between operative parameters of PCVR and PVDO in our series (Table 2), consistent with a previous study by Taylor et al. that included a mixture of syndromic and non-syndromic cases.[24,25] Compared to previous studies, we only included Apert and Crouzon syndrome cases and limited the age of surgery to increase homogeneity. The effect of syndrome type can be seen in Table 3 where transfusion volumes were higher in Apert patients than Crouzon patients undergoing SAPVE, and length of stay greater in Apert patients undergoing PVDO.

e controlled for growth between CT scans by adjusting the pre-intervention ICV measures to the post-intervention CT scan age using previously published diagnosis-specific growth curves constructed from unoperated Apert and Crouzon skull measurements.[19] The cases in our series all had an early cranial surgery and therefore may have had a lower growth potential between timepoints compared to the unoperated cases used to construct the growth curves, which would underestimate the corrected volume change. Overall, the Apert cohort ICV started close to the average unoperated Apert growth curve but became greater than this age-matched unoperated cohort after surgery (Figure 4). In comparison, the Crouzon cohort



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ICV measurements clustered more closely to the unoperated Crouzon growth curve (Figure 5).

Adjusted ICV change increases were comparable among the three groups overall (Table 5) and by syndrome (Table 6), suggesting that all techniques can achieve similar expansion however there is considerable variability. After controlling for growth, we measured 14.0% ± 12.5% increase in ICV for PCVR, 13.8% ± 5.0% for PVDO, and 17.5% ± 9.1% for SAPVE. Adjusting the pre-operative scan for growth decreases calculated volume changes compared to previously published unadjusted measures. Nowinski et al. reported unadjusted volume changes in two cases of PCVR at 13% and 24%, two cases of PVDO at 26% and 29%, and two cases of SAPVE at 18% and 25%.[26] Other groups have also reported an average unadjusted 25% volume increase after PVDO.[27,28]

Nine patients had a ventriculoperitoneal shunt (VPS) placed in our series for documented hydrocephalus, but in all but one Apert PVDO case they were placed subsequent to the posterior expansion surgery and post-operative scan. We did not find an apparent difference in the one patient who had a VPS in place at the time of PVDO. In Pfeiffer cases it is not uncommon for the VPS to be placed prior to posterior expansion, but in this Apert and Crouzon cohort VPS are less common and typically placed at an older age when they are indicated.

Although we used exclusion criteria to minimize confounding from age or diagnosis, our study has a number of limitations that prevent direct comparison. We limited our cohort to patients undergoing posterior cranial surgery at less than two years of age which pre-selected for more severe phenotypes of the two conditions. Excluded less severe Apert and Crouzon cases following the staged approach would have undergone FOA as their first surgery and not required a posterior surgery, whereas those following the expectant approach



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would have either had signs of iICP at a later age or not undergone expansion at all. Unlike previous studies on posterior expansion, we corrected for the time between CT scan measures using syndrome specific growth curves. These growth curves were compiled from unoperated Apert and Crouzon cases, and therefore may not represent the growth potential or our cohort who were a more severe phenotype subset and had a posterior surgery that may have impeded cranial growth. If growth potential was less in our groups, it would underestimate the ICV expansion that took place. Figure 4 and 5 demonstrate post-operative volumes at or greater than the unoperated growth medians which is favorable to the treatment outcome.

Our study provides a quantitative overview of two different approaches to the early treatment of syndromic synostosis – expectant and staged – as well as of three available techniques for posterior cranial expansion. It is important to emphasize that this is not intended as a direct comparison between the three surgical techniques due to the influence of team subjective decision-making, but rather a review of the outcomes of two approaches to severe syndromic synostosis requiring early posterior expansion. Our protocolized two-stage cranial expansion cohort included 10% (2/19) of Crouzon and Apert patients needing a third cranial expansion surgery for signs of iICP. In comparison, our expectant monitoring cohort that were monitored for iICP after early posterior expansion included 42% not requiring repeat expansion at last follow-up, 32% having a second expansion, and 26% a third expansion (Figures 6 and 7). Overall, this study demonstrates that either an expectant or staged approach for posterior vault expansion in severe Apert or Crouzon phenotypes using three available techniques resulted in comparable volume expansion and complication profile but resulted in two intracranial surgeries in most patients in the first decade of life.



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**Figure Legends**

Figure 1. Computerized tomography scan of an eight month old patient with Crouzon syndrome before (left) and immediately after (right) posterior cranial vault remodeling (PCVR) as part of a staged approach for intracranial volume optimization. See text for procedure details.

Figure 2. Computerized tomography scan of an eight month old patient with Apert syndrome who was treated with posterior vault distraction osteogenesis (PVDO) as part of a staged approach for intracranial volume optimization. Images are before surgery (left) and after device activation (right). See text for procedure details.

Figure 3. Computerized tomography scans of a nine month old patient with Apert syndrome who was treated with spring assisted posterior vault expansion (SAPVE) following an expectant protocol for increased intracranial pressure. Images are before surgery (left) and after spring activation (right) See text for procedure details.

Figure 4. Graph of intracranial volumes measured on computerized tomography scans of patients with Apert syndrome who underwent posterior vault expansion by the patients age at the time of the scan. Pre-expansion volumes (squares) were adjusted to an age-corrected value (diamonds) using a previously published Apert growth curve (grey curve). Dotted lines represent the estimated volume growth correction for time between scans, and solid vertical lines represent the growth-adjusted cranial volume increases from before surgery (diamonds) to after expansion (triangles).

Figure 5. Graph of intracranial volumes measured on computerized tomography scans of patients with Crouzon syndrome who underwent posterior vault expansion by the patients age at the time of the scan. Pre-expansion volumes (squares) were adjusted to an age-corrected value (diamonds) using a previously published Apert growth curve (grey curve). Dotted lines



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represent the estimated volume growth correction for time between scans, and solid vertical lines represent the growth-adjusted cranial volume increases from before surgery (diamonds) to after expansion (triangles).

Figure 6. Plot of secondary cranial surgeries that occurred in patients with Apert or Crouzon syndrome who underwent one of three types of posterior vault expansion as their first treatment. Symbols of the far left of each patient line represent the initial posterior vault surgery, with subsequent symbols depicting the secondary surgeries that took place by age. The length of each line represents the age at last follow-up for the patient to a maximum of twelve years.

Figure 7. Graph of percentage of patients following either a staged or expectant approach by number of intracranial surgeries at last follow-up up to the first decade of life. Individual patient data is depicted in figure 6. In the staged approach for treatment of visible dysmorphology, one patient only underwent a posterior cranial expansion, and the remainder had a staged posterior followed by anterior surgery with one patient having a repeat posterior surgery before the anterior surgery. Following the expectant approach for signs of increased intracranial pressure, 42% patients overall had only posterior expansion at last follow-up, 32% had a secondary cranial surgery, and 26% had a third cranial expansion.

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**Table 1: Number and percentage of Crouzon and Apert cases excluded from study by Center**

| | SCH | | GOSH | |
|---|---|---|---|---|
| | **Crouzon** | **Apert** | **Crouzon** | **Apert** |
| Total patients in clinical database | 54 | 29 | 29 | 26 |
| Excluded for first cranial surgery not being posterior* | 37 (68%) | 16 (55%) | 2 (7%) | 2 (8%) |
| Excluded for first posterior expansion age >2 years | 5 (9%) | 3 (10%) | 15 (52%) | 10 (38%) |
| Excluded for incomplete data | 3 (6%) | - | 4 (14%) | 3 (12%) |
| Included in study | 9 (17%) | 10 (34%) | 8 (27%) | 11 (42%) |

* Includes patients who had their initial surgeries elsewhere.

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**Table 2: Demographic and operative data (mean ± sd, median (IQR)) by procedure type.**

| | PCVR | PVDO | SAPVE |
|---|---|---|---|
| Number of Cases | 7 (5M, 2F) | 12 (5M, 7F) | 19 (12M, 7F) |
| Apert | 3 (1M, 2F) | 7 (2M, 5F) | 11 (7M, 4F) |
| Crouzon | 4 (4M, 0F) | 5 (3M, 2F) | 8 (5M, 3M) |
| Patient Age (months) | 7.9 ± 5.5<br>5.6 (5.2 – 9.1) | 9.1 ± 2.5<br>8.3 (7.5 – 11.1) | 11.6 ± 5.8<br>9.9 (6.5 – 17.2) |
| Total Operative Time | 3:24 ± 0:55*<br>3.22 (2:45 – 3:54) | 3:52 ± 0:34†<br>3:55 (3:27 – 4:08) | 4:34 ± 1:15†<br>4:24 (3:58 – 5:18) |
| Device Insertion | - | 2:45 ± 0:34<br>2:37 (2:20 – 3:02) | 2:38 ± 0:54<br>2:35 (2:06 – 2:56) |
| Device Removal | - | 1:07 ± 0:17*<br>1:02 (0:54 – 1:15) | 1:55 ± 0:54<br>1:39 (1:11 – 2:17) |
| Transfusion Volume (ml) | 646.3 ± 325.5*<br>566.0 (455.0 – 745.0) | 561.8 ± 319.1*<br>560.0 (393.8 – 572.5) | 234.9 ± 116.3<br>180.0 (122.0 – 250.0) |
| Length of Stay (days) | 6.0 ± 3.8<br>5.0 (3.5 – 6.5) | 8.8 ± 6.4<br>5.5 (4.0 – 15.5) | 10.7 ± 15.8<br>6.0 (4.0 – 7.0) |
| Device Insertion | - | 7.8 ± 5.9<br>5.0 (3.0 – 13.3) | 8.3 ± 15.0<br>3.0 (3.0 – 5.5) |
| Device Removal | - | 1.0 ± 1.0<br>1.0 (0.8 – 1.0) | 2.4 ± 2.9<br>1.0 (1.0 – 3.0) |
| Device in situ (months) | - | 2.9 ± 0.6*<br>2.9 (2.3 – 3.4) | 9.7 ± 6.7<br>8.7 (4.7 – 14.0) |

* Significantly different from SAPVE group ($p < 0.05$).
† Significantly different from PCVR group ($p < 0.05$).

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**Table 3: Demographic and operative data (mean ± sd, median (IQR)) by procedure type and diagnosis.**

| | Apert | | | Crouzon | | |
|---|---|---|---|---|---|---|
| | **PCVR** | **PVDO** | **SAPVE** | **PCVR** | **PVDO** | **SAPVE** |
| Number of Cases | 3 (1M, 2F) | 7 (2M, 5F) | 11 (7M, 4F) | 4 (4M, 0F) | 5 (3M, 2F) | 8 (5M, 3M) |
| Patient Age (months) | 9.8 ± 7.5<br>5.6 (5.4 – 12.0) | 9.0 ± 3.0<br>8.0 (7.1 – 11.2) | 8.1 ± 3.6**<br>7.1 (6.1 – 11.2) | 6.6 ± 4.0*<br>5.6 (4.6 – 7.5) | 9.2 ± 2.1*<br>8.5 (7.5 – 10.5) | 16.3 ± 4.9<br>17.3 (16.0 – 17.8) |
| Total Operative Time | 3:48 ± 1<br>3:56 (3:19 – 4:21) | 4:04 ± 0:38<br>4:01 (3:44 – 4:34) | 4:34 ± 1:14<br>4:24 (3:58 – 5:18) | 3:01 ± 0:45<br>2:54 (2:36 – 3:22) | 3:36 ± 0:22<br>3:46 (3:16 – 3:54) | 4:33± 1:20<br>4:28 (3:58 – 4:58) |
| Device Insertion | - | 3:00 ± 0:37<br>3:49 (3:36 – 3:37) | 2:30 ± 0:34<br>2:35 (2:08 – 2:49) | - | 2:24 ± 0:16<br>2:22 (2:13 – 2:29) | 2:50 ± 1:14<br>2:24 (2:09 – 3:07) |
| Device Removal | - | 1:04 ± 0:14*<br>1:03 (0:54 – 1:10) | 2:04 ± 0:55<br>1:46 (1:30 – 2:25) | - | 1:12 ± 0:21<br>1:02 (0:56 – 1:25) | 1:43 ± 0:55<br>1:20 (1:04 – 2:12) |
| Transfusion Volume (ml) | 811.3 ± 384.5*<br>620 (590 – 937) | 609.3 ± 412.1<br>560 (387 – 585) | 281.5 ± 122.6**<br>253 (208 – 277) | 522.5 ± 256.3*<br>455 (340 – 637) | 495.2 ± 125.6*<br>560 (456 – 560) | 168.4 ± 69.1<br>150 (125 – 230) |
| Length of Stay (days) | 4.2 ± 2.3<br>3.0 (3.0 – 5.0) | 11.9 ± 6.9<br>15.0 (5.5 – 17.5) | 9.3 ± 11.4<br>6.0 (4.0 – 7.0) | 7.3 ± 4.6<br>5.5 (4.8 – 8.0) | 4.4 ± 1.1<br>4.0 (4.0 – 5.0) | 12.6 ± 21.2<br>5.0 (4.0 – 7.3) |
| Device Insertion | - | 10.6 ± 6.4<br>13.0 (4.5 – 15.5) | 6.3 ± 8.5<br>3.0 (3.0 – 6.0) | - | 3.8 ± 1.1<br>3.0 (3.0 – 5.0) | 11.1 ± 21.4<br>3.5 (3.0 – 4.5) |
| Device Removal | - | 1.3 ± 1.3<br>1.0 (1.0 – 1.0) | 3.0 ± 3.3<br>2.0 (1.0 – 3.5) | - | 0.6 ± 0.5<br>1.0 (0.0 – 1.0) | 1.5 ± 1.9<br>1.0 (1.0 – 1.0) |
| Device in situ (months) | - | 3.0 ± 0.6<br>3.1 (2.5 – 3.4) | 8.1 ± 6.2<br>7.1 (3.5 – 10.5) | - | 2.8 ± 0.7*<br>2.5 (2.3 – 3.2) | 12.0 ± 7.0<br>12.6 (6.3 – 15.9) |

* Significantly different from diagnosis-matched SAPVE group ($p < 0.05$).
** Significantly different from surgery-matched Crouzon group ($p < 0.05$).



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**Table 4: Complications by procedure type.**

| Grade | Complication description | Number of Complications | | |
|---|---|---|---|---|
| | | **PCVR** | **PVDO** | **SAPVE** |
| 0 | No complications | 6 | 8 | 17 |
| 1 | No delay in discharge, reoperation or long-term sequelae | 1 | 1 | |
| 2 | Delay in discharge but no further operation required | | | |
| 3 | Reoperation but no long-term sequelae * | | 3 | 2 |
| 4 | Unexpected long-term deficit or neurological impairment | | | |
| 5 | Mortality | | | |

Oxford craniofacial complication scale (Paganini et al., 2019)

*** Two of PVDO and one of the SAPVE Grade 3 complications were device-related. See text for details.**

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**Table 5: Intracranial volume (ICV) changes (mean ± sd, median (IQR)) by procedure type adjusted for syndrome specific growth**

| | **PCVR** (N=7) | **PVDO** (N=12) | **SAPVE** (N=19) |
|---|---|---|---|
| **Pre-op ICV ($cm^3$)** | 842.6 ± 274.1<br>699.8 (674.0 – 972.9) | 1125.8 ± 245.3<br>1069.0 (998.4 – 1286.0) | 1067.0 ± 259.5<br>1013.9 (889.8 – 1222.1) |
| **Adjusted[1] pre-op ICV ($cm^3$)** | 1153.9 ± 253.1<br>1132.3 (1018.9 – 12221.0) | 1259.1 ± 240.2<br>1267.0 (1088.9 – 1403.2) | 1229.2 ± 224.9<br>1236.7 (1054.5 – 1345.5) |
| **Post-op ICV ($cm^3$)** | 1305.1 ± 269.2<br>1182.3 (1128.1 – 1468.7) | 1432.2 ± 277.8<br>1460.5 (1239.0 – 1581.4) | 1437.5 ± 240.9<br>1403.8 (1271.0 – 1615.1) |
| **Adjusted ICV change ($cm^3$) [2]** | 151.2 ± 129.5<br>170.7 (135.0 – 231.2) | 173.0 ± 69.7<br>151.4 (131.2 – 193.2) | 208.3 ± 100.2<br>213.7 (145.2 – 240.8) |
| **Adjusted ICV change (%) [3]** | 14.0 ± 12.5<br>16.9 (11.1 – 20.2) | 13.8 ± 5.0<br>12.9 (10.8 – 14.9) | 17.5 ± 9.1<br>14.5 (10.8 – 24.2) |
| **Significance of adjusted ICV change** | **0.02** | **<0.001** | **<0.001** |

[1] ICV on pre-operative scan adjusted for growth to time of post-operative scan using syndrome specific curves (see text for details)

[2] Adjusted ICV change = (post-op scan ICV – adjusted pre-op scan ICV)

[3] Adjusted ICV change % = (post-op scan ICV – adjusted pre-op scan ICV) / (adjusted pre-op scan ICV)

No difference of adjusted ICV change % between procedures.

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**Table 6: Intracranial volume (ICV) changes (mean ± sd, median (IQR)) by procedure type and diagnosis adjusted for syndrome specific growth**

| | Apert | | | Crouzon | | |
|---|---|---|---|---|---|---|
| | **PCVR** (N=3) | **PVDO** (N=7) | **SAPVE** (N=11) | **PCVR** (N=4) | **PVDO** (N=5) | **SAPVE** (N=8) |
| **Pre-op ICV ($cm^3$)** | 904.7 ± 369.8<br>699.8<br>(691.3 – 1015.7) | 1170.5 ± 251.4<br>1101.6<br>(1015.0 – 1291.9) | 1006.8 ± 262.7<br>941.1<br>(862.1 – 1059.1) | 796.1 ± 228.8<br>760.2<br>(642.3 -914.0) | 1063.1 ± 249.4<br>1036.3<br>(984.8 – 1172.1) | 1149.8 ± 247.0<br>1135.7<br>(971.7 – 1283.5) |
| **Adjusted[1] pre-op ICV ($cm^3$)** | 1085.2 ± 317.5<br>1302.7<br>(1164.4 – 1465.3) | 1334.8 ± 205.6<br>1349.6<br>(1211.9 – 1409.1) | 1248.3 ± 237.1<br>1239.8<br>(1118.7 – 1347.1) | 925.3 ± 356.6<br>1072.0<br>(968.0 – 1134.1) | 1248.3 ± 279.5<br>1106.6<br>(1036.0 – 1302.5) | 1312.6 ± 265.4<br>1121.9<br>(1037.2 -1312.9) |
| **Post-op ICV ($cm^3$)** | 1499.5 ± 288.9<br>1574.3 (<br>1377.4 – 1659.0) | 1503.6 ± 278.8<br>1535.3<br>(1363.3 – 1573.0) | 1483.9 ± 226.8<br>1487.6<br>(1312.4 – 1682.9) | 1159.3 ± 153.3<br>1129.0<br>(1060.8 – 1227.5) | 1332.2 ± 272.3<br>1242.3<br>(1229.2 – 1572.0) | 1373.7 ± 260.3<br>1324.3<br>(1270.3 – 1484.5) |
| **Adjusted[2] ICV change ($cm^3$)** | 180.6 ± 81.2<br>154.4<br>(135.0 – 213.0) | 164.3 ± 55.6<br>154.2<br>(143.3 – 187.8) | 241.5 ± 99.5<br>219.2<br>(173.1 – 286.7) | 129.2 ± 166.2<br>197.2<br>(98.9 – 227.5) | 185.2 ± 91.7<br>132.6<br>(127.3 – 206.3) | 162.8 ± 87.0<br>160.6<br>(92.7 – 220.8) |
| **Adjusted[3] ICV change (%)** | 14.3 ± 6.9<br>15.0<br>(11.1 – 17.9) | 12.0 ± 2.5<br>12.5<br>(11.2 – 13.6) | 20.1 ± 8.8<br>21.1<br>(13.0 – 24.8) | 13.7 ± 16.7<br>18.3<br>(10.1 – 21.9) | 16.3 ± 6.9<br>15.7<br>(11.1 – 19.9) | 13.9 ± 8.7<br>10.8<br>(7.7 – 19.0) |

[1] ICV on pre-operative scan adjusted for growth to time of post-operative scan using syndrome specific curves (see text for details)

[2] Adjusted ICV change = (post-op scan ICV – adjusted pre-op scan ICV)

[3] Adjusted ICV change % = (post-op scan ICV – adjusted pre-op scan ICV) / (adjusted pre-op scan ICV)

No difference of adjusted ICV change % between procedures or diagnosis.

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Figure 1

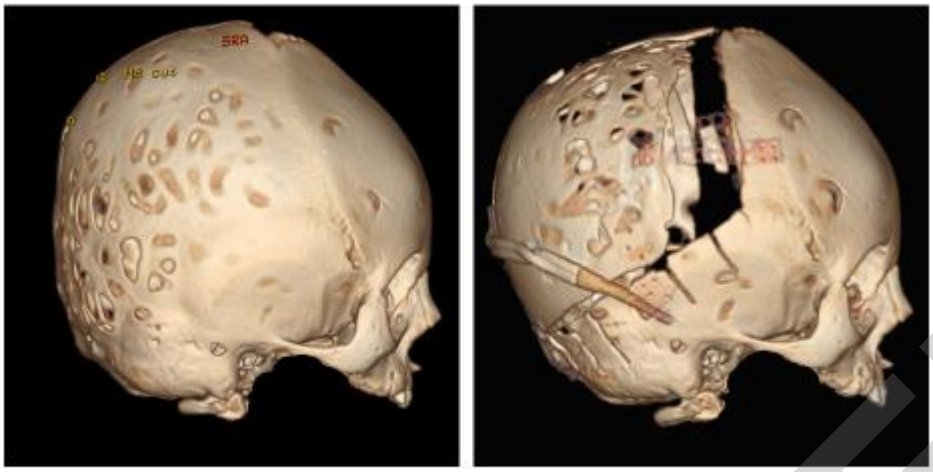

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Figure 2

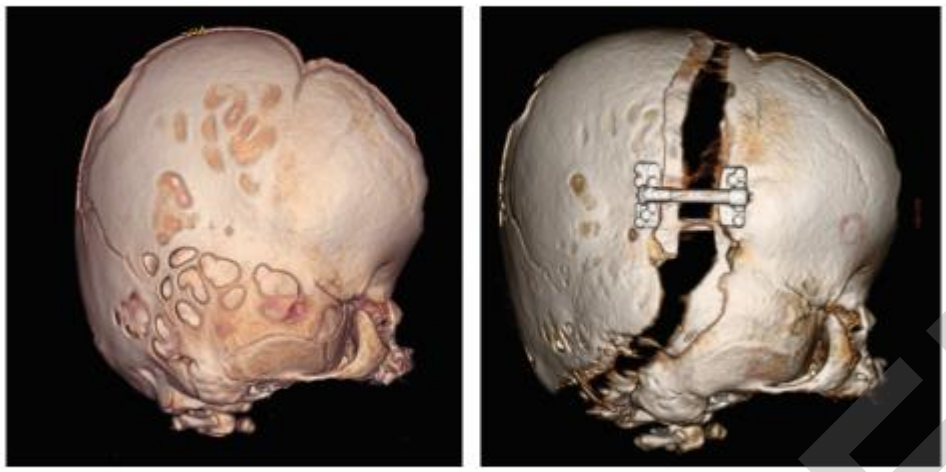

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Figure 3

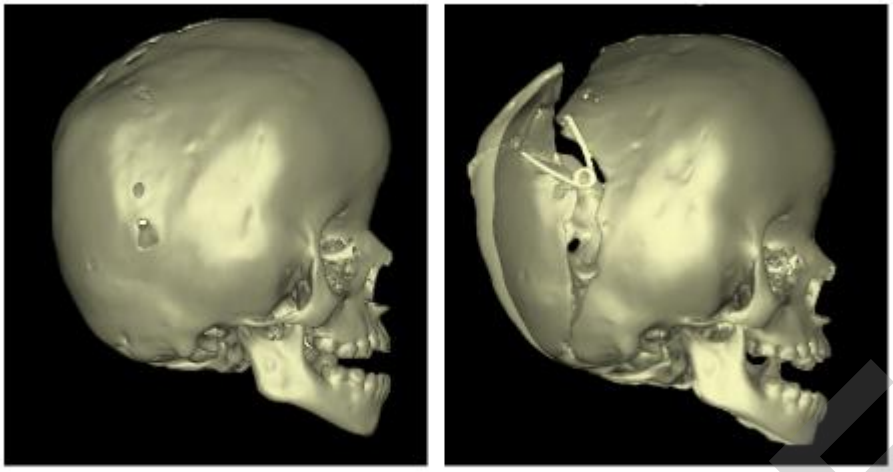

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Figure 4

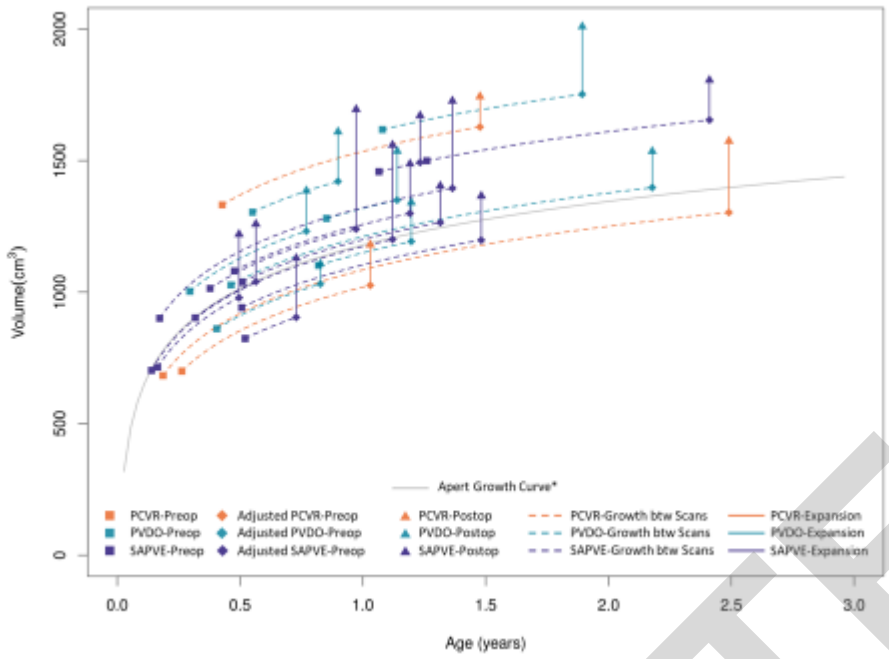


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Figure 5

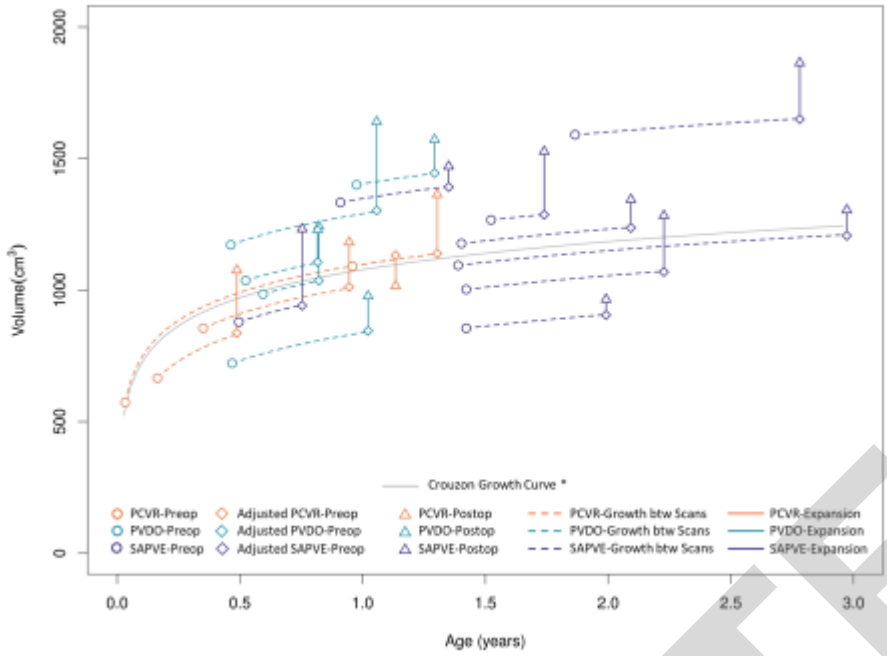


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Figure 6

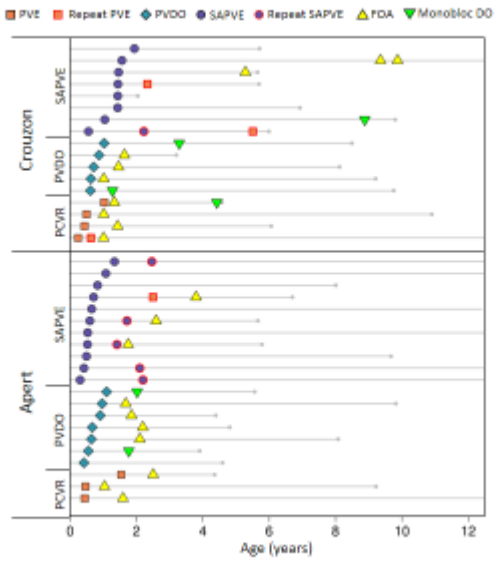


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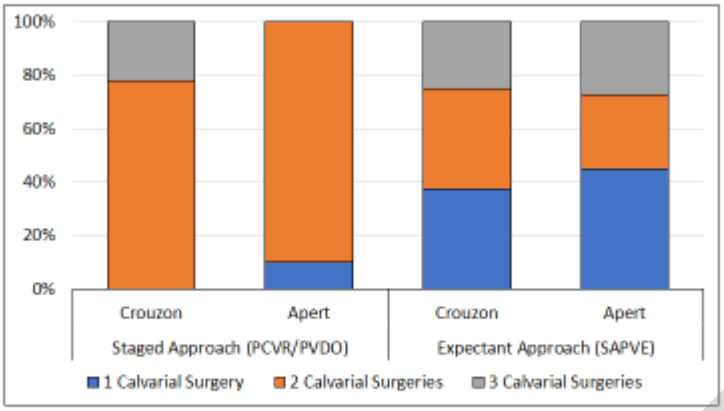
Figure 7

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