

# Successful endoscopic third ventriculostomy after craniosynostosis reconstruction with cranial vault expansion after ventriculoperitoneal shunt failure

Smit Shah, Rachana Tyagi

## ABSTRACT

**Introduction:** Syndromic craniosynostosis with growth restriction often causes increased intracranial pressure requiring shunting. Crouzon syndrome also known as branchial arch syndrome is an autosomal dominant disorder which has multiple phenotypic characteristics including proptosis, low set ears, bilateral strabismus, orofacial deformities affecting maxilla and mandible, and most importantly abnormal fusion of skull sutures also known as craniosynostosis. **Case Report:** A 17-year-old male presented with severe Crouzon syndrome and brachycephaly who had a delayed cranial vault expansion. He subsequently developed intermittent shunt malfunction, which progressed to a complete obstruction. After successful endoscopic third ventriculostomy and return to baseline function the shunt was tied off. **Conclusion:** Endoscopic third ventriculostomy (ETV) can be performed safely and effectively for shunt malfunction after cranial vault expansion in patients who may not have been candidates previously. **By using ETV in addition to known traditional techniques of**

**cranial vault expansion, patient's prognosis and postoperative recovery can be expedited.**

**Keywords:** Craniosynostosis, Crouzon syndrome, Expansion, Vault

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## INTRODUCTION

Crouzon syndrome also known as branchial arch syndrome is an autosomal dominant disorder which has multiple phenotypic characteristics including proptosis, low set ears, bilateral strabismus, orofacial deformities affecting maxilla and mandible, and most importantly abnormal fusion of skull sutures also known as craniosynostosis [1]. Numerous genetic anomalies including mutations in FGFR2 and FGFR3 have been proven to play a major role in its pathogenesis [2–4]. It is a rare disorder with an incidence of 1.6 out of every 100,000 people. Crouzon's patients typically require more orofacial reconstruction than other syndromic patients, along with intense neurosurgical intervention due to the high probability of hydrocephalus secondary to craniosynostosis and other skull base malformations [5–7].

Smit Shah<sup>1</sup>, Rachana Tyagi<sup>2</sup>

**Affiliations:** <sup>1</sup>Rutgers Robert Wood Johnson Medical School, Medical Student Year 3, New Brunswick, New Jersey, USA;

<sup>2</sup>Rutgers Robert Wood Johnson University Hospital, Pediatric Neurosurgeon, Department of Neurosurgery, New Brunswick, New Jersey, USA.

**Corresponding Author:** Smit Shah, 112 Montgomery Street, Apt 3G, Highland Park, New Jersey, USA, 08904; Email: spshah1991@gmail.com

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## CASE REPORT

A 17-year-old Filipino male presented with a significant past medical history of Crouzon syndrome was found to have clover leaf deformity, along with malignant proptosis and subluxation of globes at birth. Consequently, in 1999, he underwent bilateral cranial orbital advancement with tarsorrhaphy in addition to bifrontal craniotomy and partial removal of sphenoid wing. One month post operatively, he developed hydrocephalus for which he underwent right sided ventriculoperitoneal (VP) shunting which was revised in the year 2000, and three months later he required addition of a left VP catheter for a trapped left ventricle.

In the next few months, he developed respiratory difficulties due to a Chiari malformation extending to the C3 level, shunt failure and collapse of the bifrontal orbital advancement along with a large cervical subcutaneous muscular dural-epidural arteriovenous malformation from suboccipital area to C5. Consequently, patient underwent placement of bilateral cranial tissue expander, reprogramming of VP shunt, Chiari decompression consisting of sub occipital craniectomy and C1 to C3 laminectomy with grafting and resection of the arteriovenous malformation. The next year he underwent a nearly complete skull reconstruction for recurrent craniosynostosis (Figure 1). In 2005, he had another shunt revision, and was lost to follow-up due to insurance issues.

In 2013, he presented with respiratory failure and severe proptosis with bilateral globe subluxation with Valsalva, microcephaly and multiple bony skull defects

with no evidence of shunt malfunction. He was found to have a recurrent Chiari malformation again (Figure 2). Consequently, he underwent sub-occipital craniectomy and C1 laminectomy with expansive duraplasty and a bifrontal cranial expansion with extensive hardware for reconstruction at a specialized craniofacial center, but developed recurrent wound healing issues.

The patient presented to our system with acute neurologic compromise 10 months later, with exposed cranial mesh and enlarged ventricles (Figure 3). His shunt was externalized due to concerns for possible shunt infection with improvement in his neurologic examination and decompression of the ventricular system on repeat imaging (Figure 4). The exposed hardware was subsequently removed with a plan to address the bony defects in a delayed manner once the hydrocephalus issues were resolved. Since the cranial vault expansion, the ventricular system had expanded enough that it was felt safe to attempt an ETV (Figure 5), which was performed through a left sided approach under stereotactic guidance.

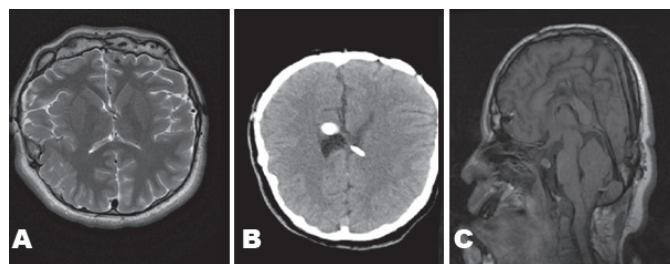


Figure 2: The patient at age 14 years. (A, B) Collapsed ventricular system, and (C) Recurrent Chiari malformation.



Figure 1: The patient at age five years demonstrating complete skull reconstruction for recurrent craniosynostosis.

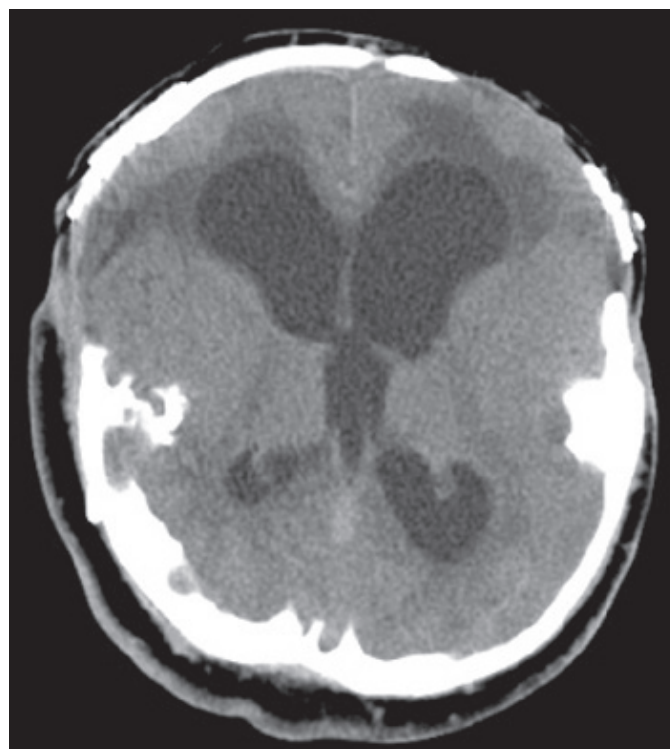


Figure 3: The patient at age 17 years 8 months demonstrating exposed cranial mesh and enlarged ventricles.

Postoperative MRI scan showed good flow through the ETV with no increase in ventricular size (Figure 6) and no change after clamping the shunt system (Figure 7). Therefore the valve was removed and the proximal right sided ventricular catheter tied off four days later, with no change in the ventricular system (Figure 8). He has continued to do well and returned to baseline neurologic function as per last follow-up 18 years 6 months.

## DISCUSSION

As mentioned before, Crouzon syndrome also known as branchial arch syndrome occurs due to abnormal embryological development of branchial arches specifically first pharyngeal arch leading to maxillary and mandibular anomalies [8–10]. Diagnosis can be made at birth by assessing phenotypical anomalies; however in overt cases CT scan and MRI scan can be used to diagnose. In patients with brain anomalies specifically increased intracranial pressure due to abnormal skull fusion, ETV can be used if conservative management fails [8, 9].

Patients with Crouzon require extensive medical and surgical management secondary to a high incidence of intracranial pressure [8, 9]. In a study performed by Abu-Sittah et al., patients nearly 50% of patients had 1–2 episodes of raised ICP and they recommended regular ophthalmologic and neurological management until age of eight years after which decision can be made about whether patients need cranial vault expansion or not depending on the degree of clinical improvement [11].

A study by Di Rocco et al. demonstrated that as the initial treatment for progressive hydrocephalus “performing an ETV may facilitate control of hydrocephalus associated with faciocraniosynostosis in select cases” in addition to mandatory radiological monitoring [12, 13]. This study included 11 pediatric patients between the age of three months and 11 years with various syndromes including Crouzon and Pfeiffer syndrome with various anatomical anomalies including cerebellar tonsil herniation, lambdoid stenosis and jugular stenosis. They demonstrated that, “ETV procedure succeeded in controlling the hydrocephalus in eight children during the first postoperative weeks, defined as disappearance of the clinical symptoms and signs of raised ICP associated with a reduction of ventricular size compared with the preoperative period. In the remaining three patients the placement of a CSF VP shunt was needed because of the persistence or prompt recurrence of the signs of raised ICP” [13]. As all the failed were performed during infancy, these patients may be candidates for a delayed ETV as in our patient. Additional larger studies assessing the efficacy of ETV in patients with craniosynostosis are required [12–14].

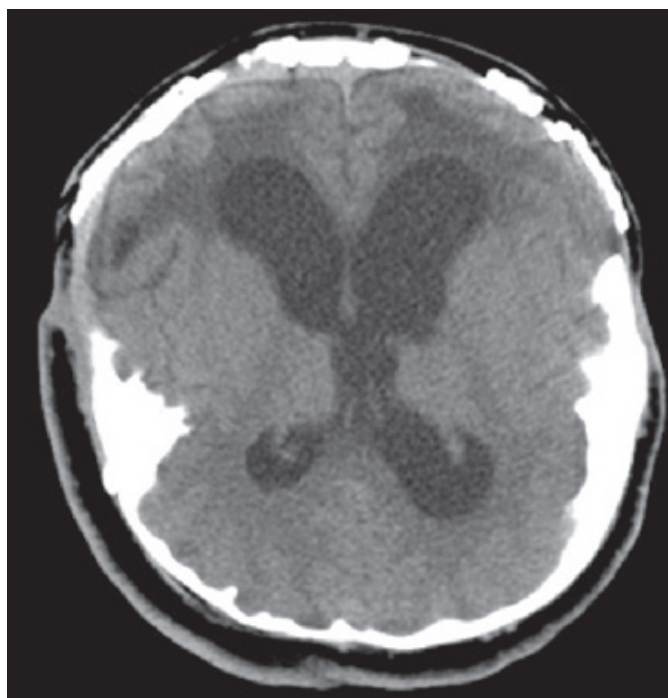


Figure 4: The patient at age 17 years 8 months showing that shunt was externalized due to concerns for possible shunt infection with improvement in neurologic exam and decompression of the ventricular system on repeat imaging.

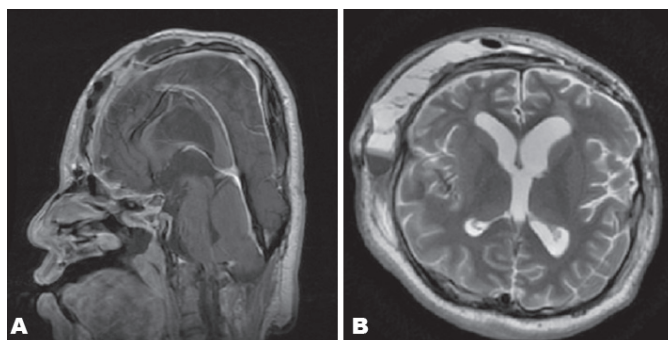


Figure 5: The patient at age 17 years 9 months. (A, B) Post cranial vault expansion, we can see expanded ventricular system which made it safe to attempt an endoscopic third ventriculostomy.

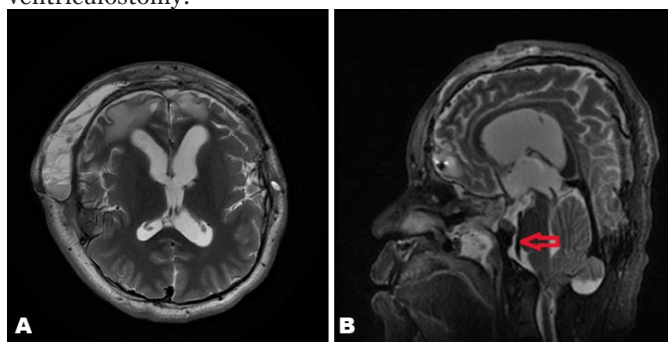


Figure 6: The patient at age 17 years 9 months. (A, B) Postoperative magnetic resonance imaging scan showing good flow through the endoscopic third ventriculostomy (arrow) with no increase in ventricular size.

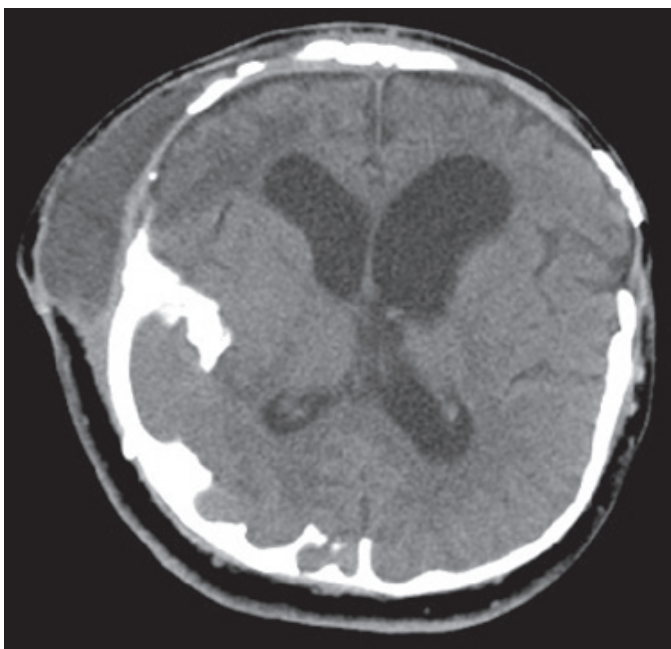


Figure 7: The patient at age 17 years 9 months showing no change in ventricular size after clamping the shunt system.

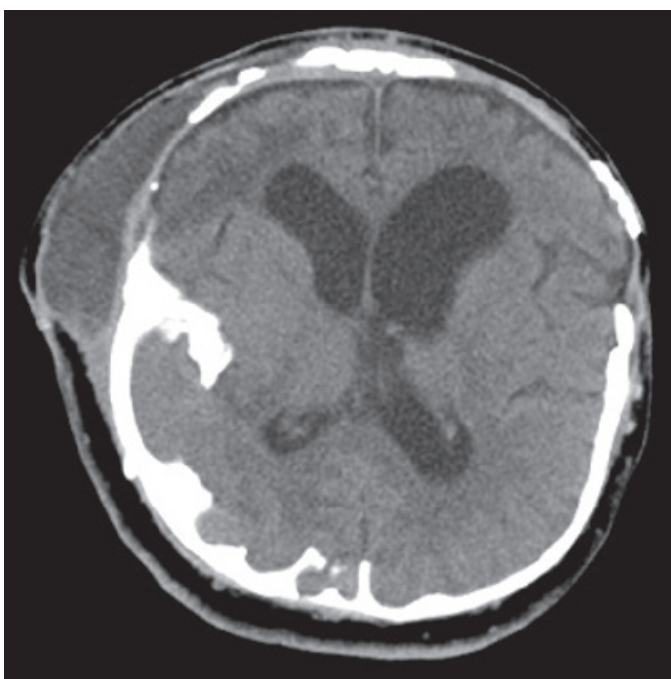


Figure 8: The patient at age 17 years 9 months showing status post valve removal and tying of the proximal right sided ventricular catheter four days later, with no change in the ventricular system.

## CONCLUSION

Endoscopic third ventriculostomy (ETV) can be performed safely and effectively for shunt malfunction after cranial vault expansion in patients who may not have been candidates previously. This article will add to the data defining possible candidates for ETV, and hopefully

increase the number of shunt-independent patients in the future. By using ETV in addition to known traditional techniques of cranial vault expansion, hopefully patient's prognosis and post-operative recovery can be expedited.

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## SUGGESTED READING

- <https://ghr.nlm.nih.gov/condition/crouzon-syndrome>
- <http://emedicine.medscape.com/article/942989-overview>
- Mallikarjunappa B, Shetty S, Gupta S, Singh J. Crouzon's syndrome: A case report from rural medical college with review of literature. *Journal International Medical Sciences Academy* 2016;29(1):30–1.

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## Author Contributions

Smit Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Rachana Tyagi – Substantial contributions to conception and design, Acquisition of data, Analysis

and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor of Submission

The corresponding author is the guarantor of submission.

## Source of Support

None

## Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

## Conflict of Interest

Authors declare no conflict of interest.

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