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Secondary Craniosynostosis and Elevated Intracranial Pressure with Functioning Shunt: An Imaging Report

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1. Abstract

A ten-year-old female pediatric patient with a history of congenital hydrocephalus and ventriculoperitoneal shunt was diagnosed with slit ventricle syndrome and severe craniosynostosis. After initial endoscopic third ventriculostomy failed, the patient underwent cranial expansion, multiple craniectomies, replacement of VP shunt and placement of an Intracranial Pressure (ICP) monitor and cranial vault reconstruction. Below are imaging studies taken preoperatively, intraoperatively, and postoperatively.

2. Introduction

A ten-year-old girl, shunted with a Codman Hakim programmable shunt on the right at approximately four months for macrocephaly and ventriculomegaly, with one proximal shunt revision at age four years, presented to the emergency room with signs and symptoms of elevated Intracranial Pressure (ICP). She had developed headaches, nausea, and vomiting. At that time, the child was taken to the operating room and a proximal shunt revision was done. The child did well for three months and then again, presented with signs and symptoms of elevated ICP. A thin cut, Computed Tomography (CT) head scan was done, and the ventricles were not enlarged (Figures 1A and 1B). A shunt tap was done, and the proximal catheter was found to be patient and the ICP was measured at 11 cm PH_20. The child was admitted to the Pediatric Intensive Care Unit (PICU) and she developed bradycardia and hypertension with heart rates as low as 40 beats per minute (bpm). A left frontal ICP monitor was placed in the PICU under sedation. While sedated, the ICP was measured between 40-60 mmHg. Once the sedation wore off, and the child's PCO_2 normalized to 35 mmHg, the ICP was measured at 8 mmHg. The child was scheduled for surgical Endoscopic Third Ventriculostomy (ETV) and drain placement, the following morning. An ETV was done on the left and a ventricular drain was placed. However, the patient could not tolerate drain clamping and required aggressive drainage of Cerebrospinal Fluid (CSF). A three dimensional (3D) reconstructed view of the head was requested and done (Figure 2). The sagittal and coronal sutures were found to be closed. A CT venogram was also done and venous sinuses were found to be small and compressed. A cranial vault expansion was scheduled. At surgery, the shunt on the right was found to be patent and the ventricular catheter and valve were functioning.

A cranial vault expansion with multiple craniectomies was performed. The ventricular catheter was replaced and a Cert as programmable valve (Codman) was used, so that reprogramming after magnetic resonance imaging would not be necessary (Figure 3A and B). Plastic surgery was consulted to perform the cranial vault remodeling. A right ICP monitor was placed for perioperative ICP management. A post-operative 3D CT scan was performed (Figure 4). The ICP's after surgery normalized and eventually, on post-operative day 2, the ICP monitor was removed. The child was discharged on post-operative day 5 and has been doing well.

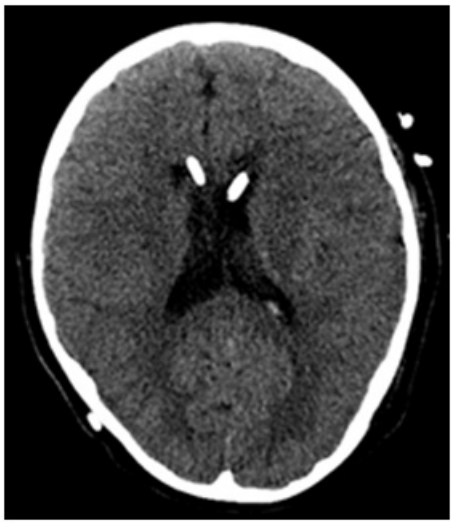


Figure 1A: CT of the brain without contrast in the axial plane after the left ETV and ventricular drain placement. Right frontal shunt catheter is shown in the right lateral ventricle and a left external ventricular drain in the left ventricle. Lateral ventricles are small.

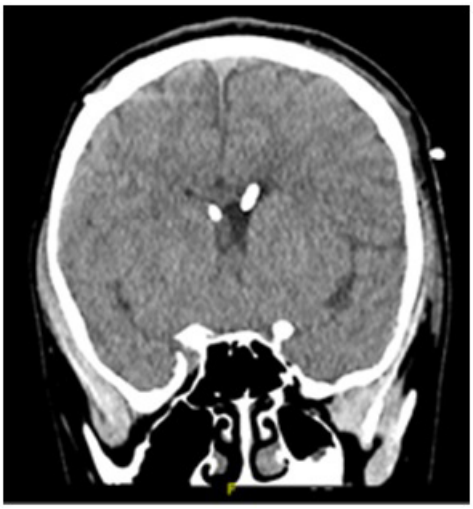


Figure 1B: CT of the brain without contrast, in the coronal plane. Right frontal shunt catheter is shown in the right lateral ventricle and a left external ventricular drain is visualized in the left ventricle. Lateral ventricles are small and third ventricle was slit-like.

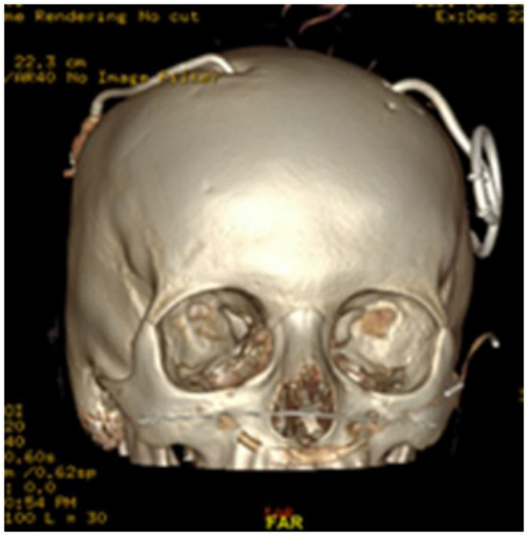
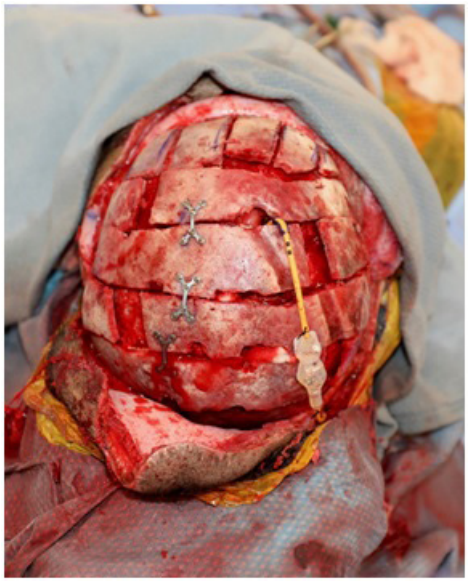


Figure 2: Three dimensional (3D) CT scan with reconstruction shows the absence of the coronal and sagittal sutures. Scan shows severe craniosynostosis and compression of all cerebral venous sinuses. Notice the shunt on the right and the external ventricular drain on the left.



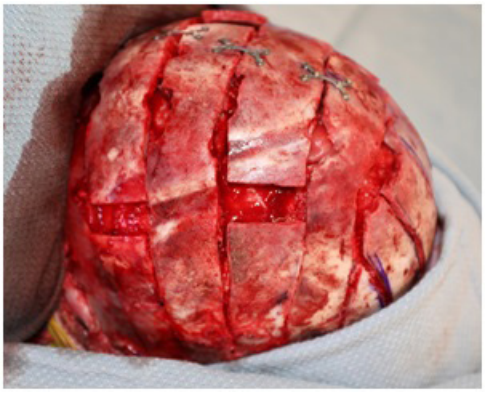


Figure 3A and 3B: Intraoperative photograph taken during cranial expansion and cranial vault reconstruction surgery. **A.** Image shows a sagittal view of the skull vault craniectomies, prior to shunt replacement. **B.** Image shows multiple strip craniectomies to open up the skull in the parietal area bilaterally in a vertex view. **Note replaced shunt on the right.**

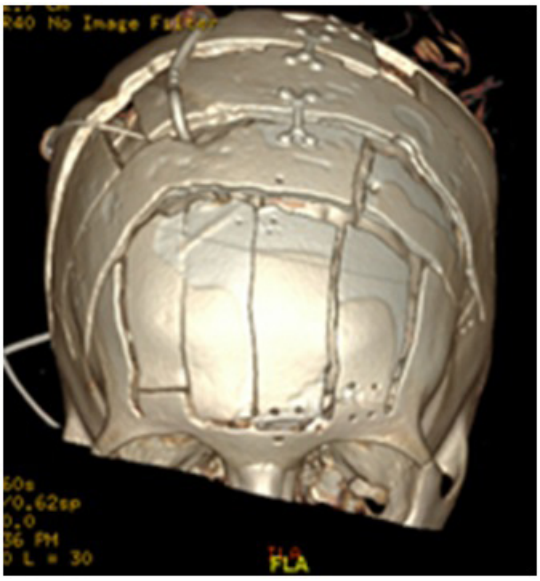


Figure 4: Postoperative CT scan with 3D reconstruction shows a cranial vault expansion with multiple frontal and bicoronal craniectomies.

3. Conclusion

For patients with recurrent shunt malfunction and small ventricles, 3D skull reconstruction views may illustrate secondary craniosynostosis, which may occur after shunt placement. Secondary craniosynostosis may occur in children with well-shunted, small ventricles and may cause elevated ICP. Luckenschadel skull appearance, with signs and symptoms of elevated ICP may be subtle signs of secondary craniosynostosis that responds well to cranial vault expansion.