The incidence of subsequent high intracranial pressure in patients undergoing early, open, and wide strip craniectomy for sagittal synostosis

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Background

The incidence of secondary bone closure following strip craniectomy for sagittal synostosis (SS) has not been systematically described. Secondary closure leading to high intracranial pressure (ICP) may cause silent papilledema, optic atrophy, and blindness.

Aim

To investigate the long-term follow-up of children who underwent early, open, and wide strip craniectomy for sagittal synostosis. Analyze the incidence of secondary closure that led to increased intracranial pressure and the subsequent need for surgical or medical intervention.

Methods

A single-center, retrospective, observational study of children who underwent early, open, and wide strip craniectomy for sagittal synostosis (SS) with a follow-up of 2 to 25 years (Av=9.1y, SD±6.2y). Surgical Technique - The patient lies down on their abdomen with head extended almost 90 degrees to the body (The 'sphinx position'). During surgery a central rectangular piece is removed using a craniotome and bone rongeur, Lateral bone cuts of about 30-40 millimeters long are performed with bone scissors with lateral reflection.

Results

The study cohort included 286 children who underwent strip craniectomy for SS between 1999-2024 with a minimum follow-up of 2 years at the age of 2-4 months (214 males). Three of 286 were diagnosed with incidental papilledema during their routine follow-up examinations (Table 1). All had documented increased intracranial pressure with formal ICP monitoring for 3 days (Codman, UK). Two of them required secondary cranial vault expansion at 1.8 and 1.9 years of age. The third patient was diagnosed with papilledema and borderline high values of ICP at the age of 2.8 years. She was treated with acetazolamide, resulting in persistent normalization of her fundoscopic exam, even after the acetazolamide treatment was discontinued.

Table 1: Patients Characteristics

Patient number	Gender	Age at surgery	Medical history	Discovery of papilledema	Symptoms	Treatment	Age at last follow-up
1	M	3 mo	Healthy	1.8 yrs	Asymptomatic	Cranial vault enlargement	3.2 yrs
2	M	3 mo	Clubfoot and Developmental delay	1.9 yrs	Asymptomatic	Cranial vault enlargement	5.3 yrs
3	F	3 mo	Healthy	2.8 yrs	Occasional headaches	Acetazolamide	6 yrs

Case 1 : A developmentally healthy boy underwent surgery for SS at 3 months of age. During a routine eye examination at age 1.8 years, papilledema was discovered clinically. Brain MRI MRV was normal. Invasive ICP monitoring (Codman, UK) over 3 days demonstrated pressure between 5-25 mmHg, including a lengthy period of elevated pressure during sleep. Therefore, he underwent cranial vault expansion. Papilledema resolved with no recurrence during a follow up of 3.2 years.

Case 2 : Sagittal craniosynostosis and scaphocephaly were noted prenatally. This developmentally delayed patient underwent surgery for SS at 3 months of age. He also had clubfoot and mild asymetry of kidney size. Genetic testing including whole exom sequencing did not detect a pathological variant. Normal fundus exam was noted prior to initial surgery. But, during a routine eye examination at age 1.9 years, papilledema was detected with increased fluid around the optic nerve on ultrasonography. Brain MRI/MRV demonstrated crowding of brain parenchyma, dilated optic nerve sheath and abnormally small transverse sinus on the right. Invasive ICP monitoring (Codman, UK) over 3 days showed pressure of 28-41 mmHg. Therefore, he underwent cranial vault expansion . Postoperatively papilledema resolved. Opic Disc appearance remained normal with a follow-up of 5.3 years. OCT RNFL images are included in figure 2.

Figure 1: MRI of Patient 2, Prior to Cranial Vault Expansion

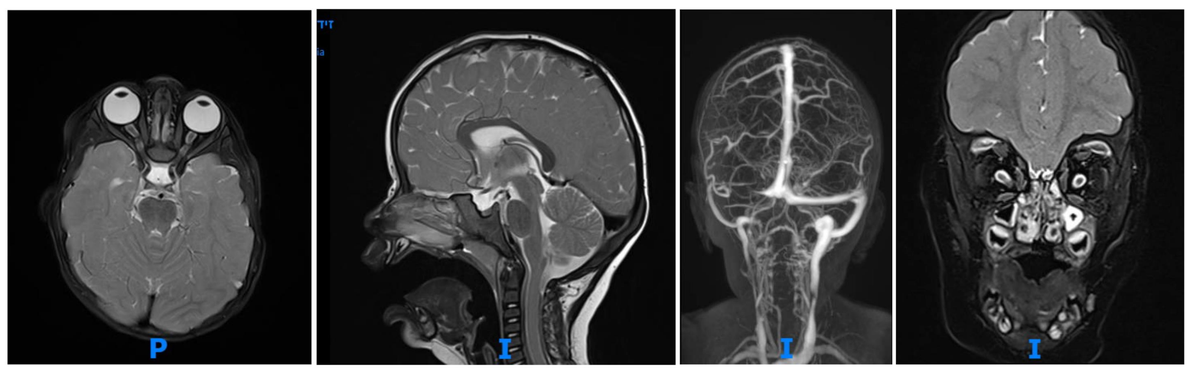
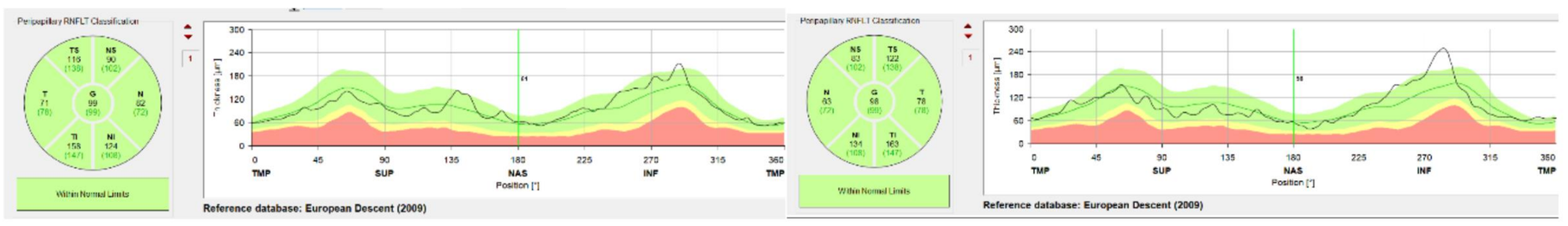


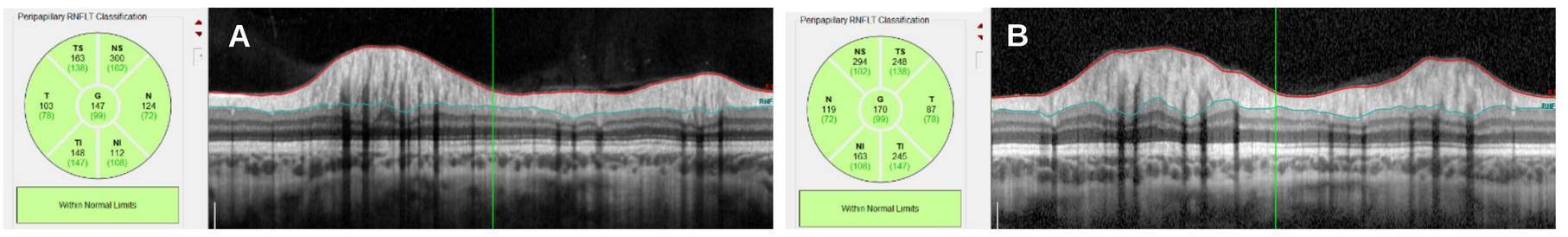
Figure 2: Case 2, Last OCT RNFL Measurements



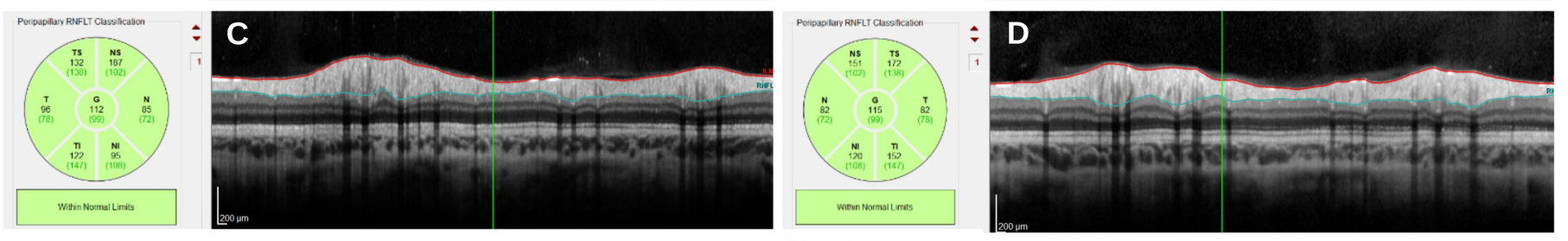
Case 3 : Healthy well developed 3 mo baby girl underwent surgery for SS. A routine eye examination at the age of 2.8 years demonstrated bilateral swollen discs. Brain MRI/MRV were normal apart from cranial vault changes and increased optic nerve sheath. Some complaints of occasional headaches were noted. Invasive ICP monitoring (Codman, UK) over 3 days showed pressure of 24-30 mmHg. Medical treatment was initiated with acetazolamide 20 mg/kg/day. It was effective in improving papilledema. She continued treatment for 15 months. The ophthalmological and neurological assessments remained normal for 2 years following cessation of acetazolamide. Figure 3 shows the improvement in RNFL measurements.

Figure 3: Patient 3 OCT images before treatment and at last follow up

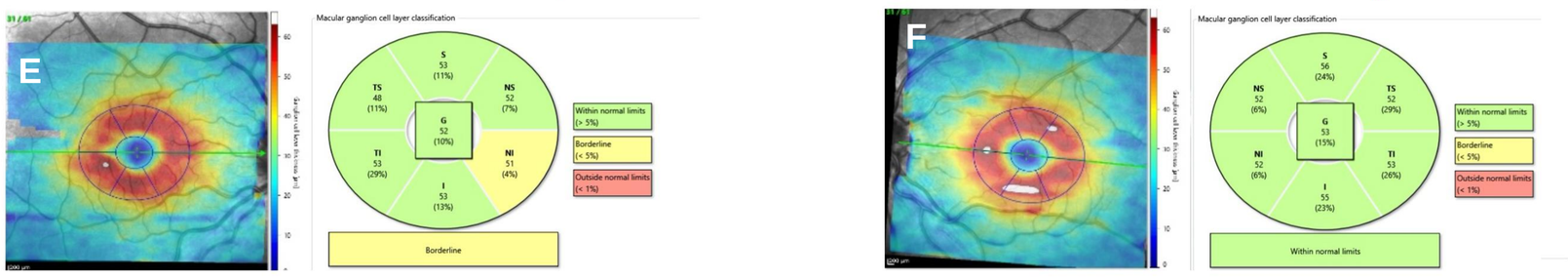
OCT RNFL images prior to treatment with acetazolamide (image A+B) with increased RNFL thickness.



OCT RNFL at last follow-up showing resolution of papilledema, a few months after cessation of treatment (C+D).



GCL is preserved (E+F).



Conclusions

This study verifies that the incidence of secondary closure is very low following early, open, and wide strip craniectomy for sagittal synostosis. However, careful clinical and ophthalmological follow-up is advised during the first few years following surgery to exclude recurrence of increased ICP and papilledema.