Outcomes of 40 Nonsyndromic Sagittal Craniosynostosis Patients as Adults: A Case-Control Study With 26 Years of Postoperative Follow-up

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Abstract

Background: While sagittal synostosis is the most common craniosynostosis, long-term follow-up of these patients is lacking.

Objective: To evaluate the results of surgical management of those patients with sagittal synostosis who attain adulthood.

Methods: An outcome study of surgically treated isolated sagittal synostosis patients operated between 1977 and 1998 was conducted at the [BLINDED] with an average follow-up time of 26.5 years. Patients’ socioeconomic situation, satisfaction with their own facial appearance and attractiveness as rated by two independent panels was evaluated and compared to controls.

Results: The self-satisfaction with the patients’ own appearance scored a mean of 75 mm on a VAS of 100 mm scale between the patients and 76 mm with the control group. The subjective satisfaction of the patients with their own appearance failed to correlate with the rating of their appearance by the panels. The panels rated the patients’ appearance to be on average 6 – 7 mm out of 100 mm VAS less attractive than the controls. Data on socioeconomic situation, including marital status, housing, education, employment of the patients and controls is presented.

Conclusion: Isolated sagittal synostosis patients treated surgically were as happy with their facial appearance as were individuals in an age and gender matched control group. Two independent panels found the patients’ appearance to be only somewhat less attractive. Analysis of the socioeconomic situation and general health of revealed that patients equaled that of controls.
Introduction

Premature fusion of the sagittal suture is the most common type of craniosynostoses, comprising 40 to 60% of all cases\(^1\). The patients have a classic scaphocephalic head shape resulting from a diminished skull height and width with compensatory anteroposterior growth\(^2\).

Surgical procedures to manage sagittal craniosynostosis have changed over the decades. Originally, the fused suture was released by making strip craniotomy lines on both sides of the suture. Later strip craniectomy of the ossified suture itself was performed\(^3,4\). Currently more extensive calvarial vault reconstructions are preferred\(^5,6\) or endoscopic strip craniectomies are done followed by postoperative molding therapy with helmets\(^3,7\). At [BLINDDED] the early operative method included two parallel strip craniotomies lateral to the midline, while a strip of bone was left over the sagittal sinus according to Lannelongue\(^4\). Later additional silicone membranes were used on the craniotomy lines\(^8\) (Fig. 1A). This technique was supplanted by suturectomy, usually supplemented with split dural interpositioning, in the early 1980’s. For dural split the outer layer of the dura was cut parallel to the sagittal sinus on both sides of it, then dissected from the deeper layer, turned over the newly cut bony edges and sutured to the periosteum (Fig. 1B)\(^9\). This method was replaced by the “H-technique” (Fig.1C)\(^5\).

From a clinical viewpoint, surgically treated sagittal craniosynostosis patients are expected to have a minimal risk of late sequelae. Clinical follow-up is usually continued no later than school age. There are several studies suggesting single suture craniosynostoses are associated with mild to moderate developmental and educational difficulties which become apparent at school age\(^10-12\). However, patients with sagittal synostosis have the best performance in neuropsychological tests\(^12\). Furthermore, all craniosynostosis may have secondary psychosocial effects. Some patients retain residual esthetic facial differences that might attract unfavourable attention from strangers and rejection from peers. Such attention may negatively influence social behaviour and result in poor school performance with difficulties in educational environments\(^13\).

When studying persons treated for severe malocclusions, it was found that the quality of life (QOL) and self-perceived esthetic satisfaction improve extensively after treatment\(^14\). These variables may be determinant of the outcomes among craniosynostoses patients, as well, in the future. However, in cases with syndromic craniosynostosis, facial differences may not influence the subjective QOL\(^15\).

The current study aimed to evaluate the esthetic outcomes in adulthood of cases treated surgically in early childhood for sagittal craniosynostosis. The evaluation of the social situation was another goal of this study.
Methods

Study patients

This was a cohort outcome study of patients and controls operated for sagittal synostosis at the [BLINDED] between 1977 and 1998. Only cases older than 18 years of age at the time of the follow-up examination were included in the study. Patients with any craniofacial syndromes or associated neurological disease were excluded. There were 171 operations for craniosynostosis during the study period. Of these, 115 patients satisfied the inclusion criteria and attempts were made to reach them (Fig. 2). Contact was made with 83 patients and 61 agreed to participate in the study. Twenty of these patients had either metopic, coronal, lambdoid or multiple suture synostosis and were not included. A single patient with both sagittal synostosis and hydrocephalus was excluded from the study. The mean age of the 40 patients (25 males, 15 females) with nonsyndromic sagittal synostosis was 27.4 years (range 18–41). The follow-up time after operation was a mean 26.5 years (range 17–37). Controls who were age and gender matched (N=40) were selected randomly from [BLINDED] Population Register Centre. Invitations were sent to both patients and controls to attend a follow-up visit by which time the subjects had completed their questionaries as requested. The facial appearance of the patients was examined during the control visit which also included evaluation of their head shape, the scar, and palpation of the head. Standardized photographs were obtained (Fig. 3). The follow-up visits took place between November 2014 and February 2016.

Operative treatment of the patients

Operative treatment included linear parasagittal craniotomies with silicone membrane interposition (N=9), suturectomy together with dural split (N=4), suturectomy without dural split (N=3), and various forms of H-plasty with or without barrel stave osteotomies of the temporal bone (N=24) (Fig. 1). The coronal and lambdoid sutures were kept intact in all cases. Mean age when surgery was performed was 5.7 months (ranging from 9 days up to 45 months). While 38 patients were operated only once, two cases required re-operation because of residual scaphocephalic head shapes. Of these two cases, one was first operated at age 9 days and then
re-operated at the age of 10.5 months. The other case was first operated at the age of 3 months and then re-operated at age 2 years.

**Esthetic evaluation using photographs**

Long-term esthetic evaluation of the clinical outcome was the major goal of the clinical evaluation. Facial esthetic appearance was rated by two independent panels. The panel members did not know which of the evaluated persons had undergone surgery and who were controls. One panel consisted of either orthodontic residents or consultants (1 male and 3 females) and one (female) orthognathic surgeon. This panel was named the “dentists’ panel”. Then a second panel which consisted of lay persons had no health care education (1 male and 3 females) were called the “lay panel”.

Panelist evaluators were presented with a series of randomly ordered slides with photographs of the patients and the controls. Each slide consisted of four images in standardized projections comprising: anterior view smiling and not smiling, with lateral and oblique views as well (Fig. 3). Slides were displayed for exactly ten seconds, in order to acquire the immediate initial panelist impression. A 100 mm VAS scale with 0 mm as least attractive and 100 mm representing most attractive was used to determine the cosmetic outcomes.

**Self-reported questionnaire**

The self-reported questionnaire contained questions regarding the participants’ education level, housing status, marital or relationship status, employment status, general health with attention to their history of headaches and the presence of mental disturbances. The education level was classified into three groups: no professional education, professional secondary education and professional tertiary education. The first group included those with no more than secondary non-vocational education accomplished. The second group included participants who had completed lower level vocational education, or any academic education with the degree below Bachelor. The third group included those with higher level vocational education or academic education of at least Bachelor level.

Participants were also directed to express their subjective satisfaction with their own facial esthetics using a 100 mm VAS. The question was: “How satisfied are you with your current facial appearance?” A VAS of 0 mm referred to “very unsatisfied” whereas a VAS of 100 mm
corresponded to “very satisfied”. Another question was “is there something that bothers you in your facial appearance?” The patients were also asked whether their scar bothered them.

**Statistical analysis**

Analyses were performed between the matched case-control pairs using Paired Samples T-test and McNemar tests. When comparing the difference between the groups, Pearson Chi-square tests and Fischer exact test were used.

The degree of the panels’ inter-observer reliability was studied using intraclass correlation coefficient both between the panels and also inside the panels amongst the panel members. When comparing two panels the reliability coefficient was almost perfect (0.857) when the patient groups were rated and substantial (0.777) for control group, and 0.840 when whole group of participants rating was analyzed. The reliability within the two panels among panel members was moderate being \( p=0.573 \) in the case of the dentists panel and \( p=0.555 \) with the lay panel. Statistical significances were set at \( p<0.05 \). All statistical analysis were performed using the commercially available SPSS software for Windows 14.0 (Chicago, Illinois, USA).

**Ethical aspects**

The study was conducted in accordance with the Helsinki declaration. The study was reviewed and accepted by the Ethical Review Board of the [BLINDED] Hospital District (number 86/2013). Both the patients and their controls signed informed consent forms.

**Results**

**Esthetic outcome according to panels evaluation**

In the case of of facial appearance evaluation by the dentists panel, the patients received lower ratings than their controls (VAS 62 vs. 69, \( p=0.002 \)). The lay panel found a similar difference with lower ratings for patients than for controls (VAS 60 vs. 66, \( p=0.011 \)) (Table 1, Fig. 4). The correlation of age of the study patients with the rating of the facial appearance by panels was not significant.
Subjective facial appearance satisfaction

The subjective grade of the patients’ own satisfaction with facial appearance compared to controls ($p=0.662$) was not significant. The mean VAS score was 75 mm for the patients and 76 mm with controls (Table 1).

Subjective satisfaction with the patients’ own appearance did not correlate with the rating by the panels (being $p=0.775$ with the dentists panel compared to $p=0.396$ for the lay panel).

When the patients and controls were asked: “Is there something that bothers you in your facial appearance (other than the scar)?” 13 patients (32.5%) and 11 controls (27.5%) answered “Yes” (McNemar $p=0.804$). Those factors which were reported to have disturbed study participants regarding their facial appearance are summarized in the Table 2. Answers for two patients were not available.

The effect of surgical technique on the outcomes

Differences between older and newer cranioplasty techniques (Table 3) regarding the mean patient age at operation and age at follow-up visit were statistically significant ($p<0.001$).

However, there was no association with operative techniques and subjective satisfaction with one’s own appearance ($p=0.801$) or the evaluation of the panels ($p=0.671$ and $p=0.922$) were noted.

The postoperative scar

Every patient who had a bothersome postoperative scar (N=4), had been operated with the H-cranioplasty using a bicoronal skin incision. One re-operated patient had a painful scar on palpation where there was an area of incomplete ossification, but the patient was satisfied with the scar appearance. The scars were visible on photographs in 6 cases, but only one of these patients found the scar to be bothersome.

General somatic health

Nine patients and 15 controls reported having migraine. Other varieties of occasional headaches were mentioned by 23 patients and 22 controls. A total of 32 of patients and 28 of controls reported having no other medical concerns. Not one case reported a history of epilepsy. Eleven (27.5%) patients and 8 (20%) controls revealed that they have or have had a history of mental
health problems. Their satisfaction with appearance did not differ from the other participants in their groups. None of these variables significant differences (McNemar $p>0.05$).

**Family and socioeconomic status**

There were no significant differences between either the patients and controls in their education level, housing situation, marital situation, or employment status (Tables 4 and 5). One half of the patients ($N=21$) and the controls ($N=20$) were involved in a permanent relationship. Likewise the same number of the patients and their controls had children with an average of 1.7 children per family among patients and 2 children per family among controls. Likewise the same number of persons in each group lived in privately owned apartments, not as tenants (Table 4).

**Discussion**

The major observation in this study was that after a mean of 26.5 years following surgery for scaphocephaly, patients were equally satisfied with their appearance as the controls. The same applies to their general physical health. Their family status was also equal.

The predominance of males among sagittal synostosis patients is in agreement with previous studies, though the ratio of males to females was 1.7:1 in our study population, which was lower than the 2.6-3.8:1 ratio from previous studies\(^1,16\). Demographic data in controls such as living with parents, being unemployed or education was in accordance with average numbers for the [BLINDED] population, confirming that the control cohort was representative\(^17\).

**Panels evaluation of appearance**

Both panels rated the patients’ appearance to be worse than the appearance of the controls. The difference appeared to be statistically significant, but was only 7 mm out of a 100 mm of VAS as rated by the panel of dentists and 6 mm as rated by the panel of lay persons (Table 1, Fig. 4).

Few studies evaluate craniosynostosis surgical outcomes from photographs using independent panels of evaluators\(^18-20\). Though this method is widely used in orthodontic and cleft-lip-palate surgery evaluations\(^14,21\) it is just being introduced into the craniosynostosis surgical community. Since different follow-up times, imaging sets, scoring systems were used and different size and background of panelists in every study so far, the results cannot be directly compared.

A lay panel was used for the first time in the assessment of operative results following craniosynostosis surgery from photographs in a study by Metzler et al. They found no significant
difference between the esthetic scores among panels of professionals and lay persons when full-size facial images were evaluated. This finding is in agreement with our study. But the score used in the study by Metzler had only 3 grades with no patients rated to be in the worst grade 3 when full-size images were evaluated. This finding speaks for the need to use more precise tools and scores able to detect changes of smaller magnitude. Esthetic evaluation of operative results is highly subjective and possible influenced by many factors. This probably explains why in spite of almost perfect agreement between the panels evaluations, there was only moderate agreement between individual panellists. Studies are needed to define standards for image set, scores to be used, definition of questions, panel size and panel constituents.

The difference in magnitude between panel evaluation of patients and controls of less than 10 mm in the VAS raises the question of minimally clinically important difference (MCID). Though this difference is statistically significant, it is less than 10% and it could be of no clinical importance. In the literature no standard is reported for determining MCID when VAS scales are used in the evaluating esthetic surgical outcomes.

**Self-perceived satisfaction with facial appearance**

Patients appeared to be just as satisfied with their facial appearance as compared to the control group. The appearance of the postoperative scar bothered only few patients (10%). All these patients graded their satisfaction with their general appearance below average as well. Also all four of these patients were operated using a bicoronal incision with the H-cranioplasty technique. While the sagittal direction of skin incision is not applicable when performing modern extended calvarial remodelling surgery, this direction of the scar seems to bear less cosmetic concern. Surprisingly the visibility of the scar seemed not to correlate with the patients’ satisfaction with own appearance.

There was no significant difference between the groups regarding the issue of unsatisfactory facial appearance, with teeth, shape of head or face being the most common subjects to complain about in both patients and controls (Table 2). The majority of patients’ complaints (8 out of 13) were not related to the craniosynostosis itself.

No correlation was revealed between their self-perceived satisfaction in facial appearance and the results of panels’ evaluations neither in patients, nor was this noted in the control group. Contrasting with the current findings, when self-perceived satisfaction with appearance was compared to panel evaluations of patients presenting for severe malocclusion treatments or other esthetic facial surgery, there was usually some moderate correlation found. Unlike those
studies, our patients were not independently seeking treatment, but were invited for a follow-up visit years after treatment. Our findings were in agreement with previously published studies on body image of persons with visible differences (e.g. other craniofacial malformations, amputations of limbs or burn injuries), where the degree of disfigurement does not predict the magnitude of negative body image or how people experienced their bodies\textsuperscript{24}.

The effect of operative technique on the outcomes

The clinical milieu where these patients were treated has changed during the years. Modern surgical techniques have been developed and the medical community and public in general have become more aware of craniosynostoses, leading to a decrease in delayed diagnoses\textsuperscript{1}. This trend is demonstrated in our data. Also all 3 of the cases in our study group that were operated after 1 year of age, were operated in the 1970-s. Older techniques of linear craniotomy and suturectomy were abandoned in 1985, and replaced by modifications of the H-technique. Accordingly, the direction of the scar was changed too from sagittal to bicoronal. Thus the patients who were operated with new methods were about 10 years younger at follow-up visits. It could be argued that this could influence the results of self-evaluation or the panels’ evaluation. However, the authors found no correlation of any esthetic or self-evaluation parameters with age in this study. Surprisingly the authors found no differences in outcomes between the different operative techniques (Table 3). However, lacking data on the preoperative severity of the deformities the authors do not know how decision making in favor of surgery changed over the years. It is possible that patients operated 30 – 40 years ago with this method had less significant degrees of scaphocephalic deformity preoperatively than the patients that were operated during 1990-s. Since this study employs historical cohorts with different operative methods used during certain time periods independently of the severity of scaphocephalic deformation, there is likely no patient selection bias when choosing operative method.

Somatic and mental health

Isolated sagittal synostosis has usually no influence on somatic health. This was confirmed by the current findings. The authors also found no correlation with surgically treated sagittal synostosis and the self-reported prevalence of headaches or mental problems. Having a history of mental problems did not appear to influence any study participant’s satisfaction with their facial appearance.
Family and socioeconomic situation

Having surgery for sagittal craniosynostosis did not have an influence on a patients’ level of education, status in employment, the finding of a life partner or the having of children later in life (Tables 4 and 5).

According to the European Union statistics data, 4% the proportion of young people aged 25 – 34 years who still live with their parents is 4% in [BLINDED]. In the present study 2 persons among the controls (5 %) and 5 of the patients (12.5%) still continued to live together with their parents\(^\text{17}\). This difference between the patients and controls was not significant, and could be explained by a relatively small sized population and the young age, since none of those still living with parents was over 23 years of age. A total of 42.7% of persons aged 30 – 34 years in [BLINDED] had a tertiary level of education\(^\text{17}\). Notably 30% of patients and corresponding 32.5% of controls had professional tertiary level education. However, the participants of this study were younger than the reference population and some were still engaged in studying. There were 5 unemployed patients (12.5%) and 3 controls (7.5%). The general level of unemployment in the year 2015 in [BLINDED] ranged from 10 – 15% in persons with ages ranging from 15 – 74 years\(^\text{17}\). Thus the study results were in agreement with these numbers.

While there is a growing amount of data on late follow-up of syndromic cases, data on adult single suture synostoses is still scarce. Our study had intended to fill this gap and to give craniofacial surgeons as well as patients and their families more accurate information on expected long-term results of the surgery, and that also from patients’ point of view.

Conclusions

Adult patients treated for sagittal craniosynostosis in early childhood managed as well in adult life as controls. Patients had equal headache frequencies, rates of mental health problems or issues with general health as the control group. Patients were also as satisfied with their own facial appearance as were their age and gender matched control groups.

Both panels of lay personnel and dentists found the patients’ appearance to be slightly less attractive than controls, but this difference was slight, being less than 10 mm on a 100 mm VAS. There was no correlation between esthetic self-satisfaction and third person’s opinion regarding facial appearance.

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References:


**Figure legends:**

**Figure 1.** Different techniques used for treatment of sagittal craniosynostosis in the [BLINDED]. A. The Lannelongue and Ingraham suture “unlocking” technique with and without silicone
membrane interposition. B. Lane strip craniectomy with and without dural splitting. C. “H” cranioplasty with and without additional cuts to the temporal bone.

**Figure 2.** Flow chart of the patients included into the study.

**Figure 3.** An example of the slide with photographs as presented for the evaluation of esthetic results of the surgery by panelists. An image of a healthy person not included in the study is present with his consent. On the slides presented to the panels the eyes of the study persons were not blinded.

**Figure 4.** Ratings of facial appearance as evaluated by the panels and subjects self-satisfaction with facial appearance on the 100 mm Visual Analogue scale.