

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

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28

29 **Abstract**

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

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

34 **Methods:** An outcome study of surgically treated isolated sagittal synostosis patients operated  
35 between 1977 and 1998 was conducted at the [BLINDED] with an average follow-up time of  
36 26.5 years. Patients' socioeconomic situation, satisfaction with their own facial appearance and  
37 attractiveness as rated by two independent panels was evaluated and compared to controls.

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

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

## 49 **Introduction**

50 Premature fusion of the sagittal suture is the most common type of craniosynostoses, comprising  
51 40 to 60% of all cases<sup>1</sup>. The patients have a classic scaphocephalic head shape resulting from a  
52 diminished skull height and width with compensatory anteroposterior growth<sup>2</sup>.

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

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

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

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

82

## 83 **Methods**

### 84 **Study patients**

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

102

### 103 **Operative treatment of the patients**

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

111 re-operated at the age of 10.5 months. The other case was first operated at the age of 3 months  
112 and then re-operated at age 2 years.

113

## 114 **Esthetic evaluation using photographs**

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

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

## 128 **Self-reported questionnaire**

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

138 Participants were also directed to express their subjective satisfaction with their own facial  
139 esthetics using a 100 mm VAS. The question was: “How satisfied are you with your current facial  
140 appearance?” A VAS of 0 mm referred to “very unsatisfied” whereas a VAS of 100 mm

141 corresponded to “very satisfied”. Another question was “is there something that bothers you in  
142 your facial appearance?” The patients were also asked whether their scar bothered them.

## 143 **Statistical analysis**

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

147 The degree of the panels’ inter-observer reliability was studied using intraclass correlation  
148 coefficient both between the panels and also inside the panels amongst the panel members. When  
149 comparing two panels the reliability coefficient was almost perfect (0.857) when the patient  
150 groups were rated and substantial (0.777) for control group, and 0.840 when whole group of  
151 participants rating was analyzed. The reliability within the two panels among panel members  
152 was moderate being  $p=0.573$  in the case of the dentists panel and  $p=0.555$  with the lay panel.

153 Statistical significances were set at  $p<0.05$ . All statistical analysis were performed using the  
154 commercially available SPSS software for Windows 14.0 (Chicago, Illinois, USA).

## 155 **Ethical aspects**

156 The study was conducted in accordance with the Helsinki declaration. The study was reviewed  
157 and accepted by the Ethical Review Board of the [BLINDED] Hospital District (number  
158 86/2013). Both the patients and their controls signed informed consent forms.

## 159 **Results**

### 160 **Esthetic outcome according to panels evaluation**

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

## 166 **Subjective facial appearance satisfaction**

167 The subjective grade of the patients' own satisfaction with facial appearance compared to  
168 controls ( $p=0.662$ ) was not significant. The mean VAS score was 75 mm for the the patients and  
169 76 mm with controls (Table 1).

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

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

## 177 **The effect of surgical technique on the outcomes**

178 Differences between older and newer cranioplasty techniques (Table 3) regarding the mean  
179 patient age at operation and age at follow-up visit were statistically significant ( $p<0.001$ ).  
180 However, there was no association with operative techniques and subjective satisfaction with  
181 one's own appearance ( $p=0.801$ ) or the evaluation of the panels ( $p=0.671$  and  $p=0.922$ ) were  
182 noted.

## 183 **The postoperative scar**

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

## 189 **General somatic health**

190 Nine patients and 15 controls reported having migraine. Other varieties of occasional headaches  
191 were mentioned by 23 patients and 22 controls. A total of 32 of patients and 28 of controls  
192 reported having no other medical concerns. Not one case reported a history of epilepsy. Eleven  
193 (27.5%) patients and 8 (20%) controls revealed that they have or have had a history of mental

194 health problems. Their satisfaction with appearance did not differ from the other participants in  
195 their groups. None of these variables significant differences (McNemar  $p>0.05$ ).

## 196 **Family and socioeconomic status**

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

## 203 **Discussion**

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

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

## 212 **Panels evaluation of appearance**

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

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

221 A lay panel was used for the first time in the assessment of operative results following  
222 craniosynostosis surgery from photographs in a study by Metzler et al. They found no significant



223 difference between the esthetic scores among panels of professionals and lay persons when full-  
224 size facial images were evaluated<sup>20</sup>. This finding is in agreement with our study. But the score  
225 used in the study by Metzler had only 3 grades with no patients rated to be in the worst grade 3  
226 when full-size images were evaluated. This finding speaks for the need to use more precise tools  
227 and scores able to detect changes of smaller magnitude. Esthetic evaluation of operative results  
228 is highly subjective and possible influenced by many factors. This probably explains why in spite  
229 of almost perfect agreement between the panels evaluations, there was only moderate agreement  
230 between individual panellists. Studies are needed to define standards for image set, scores to be  
231 used, definition of questions, panel size and panel constituents.

232 The difference in magnitude between panel evaluation of patients and controls of less than 10  
233 mm in the VAS raises the question of minimally clinically important difference (MCID) <sup>22</sup>.  
234 Though this difference is statistically significant, it is less than 10% and it could be of no clinical  
235 importance. In the literature no standard is reported for determining MCID when VAS scales are  
236 used in the evaluating esthetic surgical outcomes.

### 237 **Self-perceived satisfaction with facial appearance**

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

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

250 No correlation was revealed between their self-perceived satisfaction in facial appearance and  
251 the results of panels' evaluations neither in patients, nor was this noted in the control group.  
252 Contrasting with the current findings, when self-perceived satisfaction with appearance was  
253 compared to panel evaluations of patients presenting for severe malocclusion treatments or other  
254 esthetic facial surgery, there was usually some moderate correlation found<sup>14,23</sup>. Unlike those

255 studies, our patients were not independently seeking treatment, but were invited for a follow-up  
256 visit years after treatment. Our findings were in agreement with previously published studies on  
257 body image of persons with visible differences (e.g. other craniofacial malformations,  
258 amputations of limbs or burn injuries), where the degree of disfigurement does not predict the  
259 magnitude of negative body image or how people experienced their bodies<sup>24</sup>.

## 260 **The effect of operative technique on the outcomes**

261 The clinical milieu where these patients were treated has changed during the years. Modern  
262 surgical techniques have been developed and the medical community and public in general have  
263 become more aware of craniosynostoses, leading to a decrease in delayed diagnoses<sup>1</sup>. This trend  
264 is demonstrated in our data. Also all 3 of the cases in our study group that were operated after 1  
265 year of age, were operated in the 1970-s.

266 Older techniques of linear craniotomy and suturectomy were abandoned in 1985, and replaced  
267 by modifications of the H-technique. Accordingly, the direction of the scar was changed too from  
268 sagittal to bicoronal. Thus the patients who were operated with new methods were about 10 years  
269 younger at follow-up visits. It could be argued that this could influence the results of self-  
270 evaluation or the panels' evaluation. However, the authors found no correlation of any esthetic  
271 or self-evaluation parameters with age in this study.

272 Surprisingly the authors found no differences in outcomes between the different operative  
273 techniques (Table 3). However, lacking data on the preoperative severity of the deformities the  
274 authors do not know how decision making in favor of surgery changed over the years. It is  
275 possible that patients operated 30 – 40 years ago with this method had less significant degrees  
276 of scaphocephalic deformity preoperatively than the patients that were operated during 1990-s.  
277 Since this study employs historical cohorts with different operative methods used during certain  
278 time periods independently of the severity of scaphocephalic deformation, there is likely no  
279 patient selection bias when choosing operative method.

## 280 **Somatic and mental health**

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

286 **Family and socioeconomic situation**

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

290 According to the European Union statistics data, 4% the proportion of young people aged 25 –  
291 34 years who still live with their parents is 4% in [BLINDED]. In the present study 2 persons  
292 among the controls (5 %) and 5 of the patients (12.5%) still continued to live together with their  
293 parents<sup>17</sup>. This difference between the patients and controls was not significant, and could be  
294 explained by a relatively small sized population and the young age, since none of those still  
295 living with parents was over 23 years of age. A total of 42.7% of persons aged 30 – 34 years in  
296 [BLINDED] had a tertiary level of education<sup>17</sup>. Notably 30% of patients and corresponding  
297 32.5% of controls had professional tertiary level education. However, the participants of this  
298 study were younger than the reference population and some were still engaged in studying. There  
299 were 5 unemployed patients (12.5%) and 3 controls (7.5%). The general level of unemployment  
300 in the year 2015 in [BLINDED] ranged from 10 – 15% in persons with ages ranging from 15 –  
301 74 years<sup>17</sup>. Thus the study results were in agreement with these numbers.

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

306

307 **Conclusions**

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

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

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374 special populations. *Body Image*. 2015;14:168-176.

375

## 376 **Figure legends:**

377

378 **Figure 1.** Different techniques used for treatment of sagittal craniosynostosis in the [BLINDED].  
379 A. The Lannelongue and Ingraham suture “unlocking” technique with and without silicone

380 membrane interposition. B. Lane strip craniectomy with and without dural splitting. C. “H”  
381 cranioplasty with and without additional cuts to the temporal bone.

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

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

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

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