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The incidence of craniosynostosis in the Netherlands, 1997–2007☆

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Summary *Objects:* The first aim of this study was to determine the incidence of craniosynostosis, metopic synostosis and sagittal synostosis in the Netherlands from 1997 to 2007. The second aim was to study whether a shift in the proportion of different subtypes of craniosynostosis had taken place in the Netherlands during the study period.
Methods: Patients of all six units performing craniofacial procedures in the Netherlands that met the inclusion criteria participated in the study ($n = 927$). A logistic regression in a weight sample was used to assess the differences in both the proportion and the incidence of different subtypes of craniosynostosis each year. The angle of the metopic ridge was compared using a regression analysis.
Results: The incidence of craniosynostosis increased from 2.6 per 10 000 live births in 1997 to 6.4 in 2007. The incidence of metopic synostosis showed a significant increase of 6% each year ($p = 0.029$). We observed a significant change in the proportion of metopic synostosis, with a mean percentage of 20% during 1997–2000 and 27% during 2001–2007 ($p = 0.046$). The incidence of sagittal synostosis annually increased by 3% ($p = 0.89$). We did not find a significant change regarding the proportion of sagittal synostosis during 2000–2001. No significant difference in the severity of metopic synostosis was found between 1997–2000 and 2001–2004.
Conclusion: The incidence of craniosynostosis, metopic synostosis and sagittal synostosis is 6.4; 1.9; and 2.8 per 10,000 live births, respectively. Both the incidence and the proportion of metopic synostosis have significantly increased over the study period, concluding that metopic synostosis is on the rise.

Premature closure of the sagittal suture results in a skull malformation known as scaphocephaly. The skull shape is elongated and narrow with a ridge running along the sagittal suture.[1] Although sagittal synostosis is known to be the most benign form of synostosis, cognitive and behavioural delays have also been reported.[1–5] The incidence of

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sagittal synostosis is estimated at 1 in 4200–5800 live births[6] and the proportion of sagittal synostosis in the literature ranges from 22% to 68%.[1, 6–16] These percentages are compared with the total number of craniosynostosis cases found in previous studies.

Premature closure of the metopic suture leads to a skull malformation known as trigonocephaly. Typically, a wedge-shaped forehead is seen because of the supraorbital recession, which is combined with hypotelorism.[1] The volume of the anterior cranial fossa is reduced, although the total skull volume is often unrestricted.[1] There have been reports on cognitive and behavioural delays in children with metopic synostosis.[2–5,17] The incidence of metopic synostosis is estimated at 1 in 15,000 live births[18] and the proportion of metopic synostosis in the literature ranges from 3% to 27%.[1,6,7,9–16]These percentages are compared with the total number of craniosynostosis cases found in previous studies. The incidence of craniosynostosis is estimated at 3.1 to 4.8 per 10,000 live births.[14,19–21] Recent studies show an increase in the absolute number of patients with metopic and sagittal synostosis over the past decade.[1,14–16,22] Furthermore, two recent studies showed a shift in the proportion of metopic synostosis as regards single-suture synostoses.[1,15] All together, this suggests that metopic synostosis is on the rise. The first aim of this study was to determine the incidence of craniosynostosis, metopic synostosis and sagittal synostosis in the Netherlands from 1997 to 2007. The second aim was to study whether a shift in the proportion of different subtypes of craniosynostosis had taken place in the Netherlands during the study period.

Materials and methods

Patient sample

Patients of all six units performing craniofacial procedures in the Netherlands, who met the inclusion criteria (diagnosed with primary craniosynostosis and born in the Netherlands between 1 January 1997 and 31 December 2007), participated in the study.

Data were collected by consulting the Dutch Association for Cleft Palate and Craniofacial Anomalies (NVSCA) and through personal communication with craniofacial surgeons of each participating centre. Diagnosis was confirmed by physical examination and radiographic imaging (computed tomography (CT) scan). The final study sample consisted of 927 patients presenting with primary craniosynostosis. All patients included in this study were treated surgically. The patient population consisted of two craniosynostosis subgroups: a total of 757 patients with single-suture craniosynostosis (plagiocephaly, trigonocephaly and scaphocephaly) and 170 patients with complex craniosynostosis (brachycephaly; oxycephaly; cloverleaf skull; and Crouzon, Saethre-Chotzen, Apert, Cohen, Pfeiffer, Carpenter, Say–Meyer and Antley–Bixler syndromes).

Procedure and instruments

The population dynamics of the Netherlands (population rate and live birth rate) from 1 January 1997 to 31 December 2007 were obtained from Statistics Netherlands. One of the explanations for the increase of metopic synostosis might be better recognition of skull deformations. To investigate whether more patients presented with less severe forms of metopic synostosis over the last decade, we selected a random sample ($n = 76$) of patients from the Sophia Children's Hospital Rotterdam diagnosed with metopic synostosis and born between 1 January 1997 and 31 December 2004, and measured the angle of the metopic ridge. This was defined as the angle between the two lines drawn through the pterion (bilaterally) and the nasion, as described by Oi and Matsumoto in 1986 on a preoperative CT (Figure 1).[23]

Statistical analysis

First, a logistic regression in a weight sample was used to assess the differences in both the proportion and the incidence of different subtypes of craniosynostosis each year compared with the total amount of craniosynostosis cases each year. Second, to investigate whether less severe forms of metopic synostosis presented in time, we compared the angle of the metopic ridge using a linear regression analysis.

Results

In the six Craniofacial Units in the Netherlands, the total number of infants born in the study period, with diagnosed craniosynostosis, was 927 (Table 1). The absolute numbers and the proportion of craniosynostosis, metopic synostosis and sagittal synostosis are summarised in Table 1. The population dynamics of the Netherlands during the study period are described in Table 2. During the study period, the total Dutch population increased from 15,567,107 in 1997 to 16,357,992 people in 2007, while the amount of total live births decreased from 192,443 in 1997 to 181,336 in 2007 (Table 2). The sex ratio remained relatively stable between 1997 and 2007 (Table 2). Four out of six units

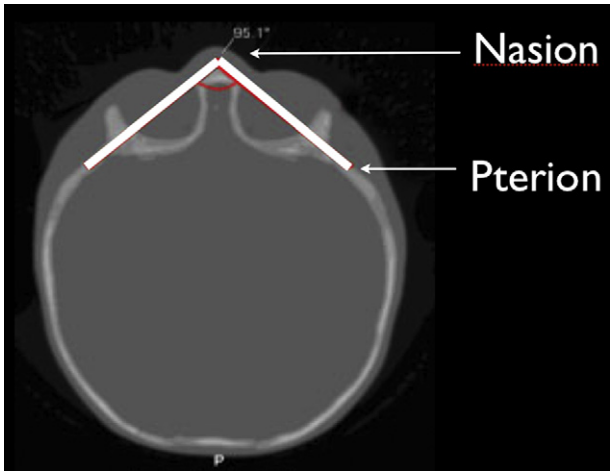

Figure 1 Measurement for the severity of metopic synostosis: the angle between the two lines drawn through pterion (bilaterally) and nasion.