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The value of anthropometrical measurements in a case of Hallermann-Streiff syndrome

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INTRODUCTION

On the long list of cranio-facial dysostoses, the Hallermann-Streiff syndrome or oculo-mandibulo-dyscephaly with hypotrichosis is one of the most uncommon. The first case was described by Aubry in 1893 under the name of "Alopécie congénitale suturale". Since then, 50 other cases have been reported. None of these reports was performed on an anthropometrical basis. In our opinion, however, the only valid way to evaluate a case of cranio-facial dysostosis is with the help of anthropometry.

From the literature follows that this syndrome is invariably characterised by seven major symptoms: dyscephaly and bird face, dental anomalies, harmonious namism, hypotrichosis, skin atrophy, bilateral cataract and micropthalmy.

ANTHROPOMETRICAL DATA

Method: The only way to perform an anthropometrical evaluation of the

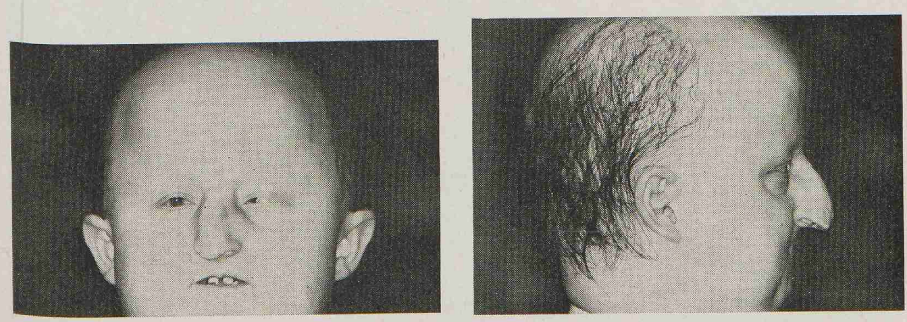

Figure 1. The patient (1.50 m, 50 kg, 30 years old).

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skull is by using standardized röntgenograms. This kind of röntgenograms can only be obtained with a cephalostat in which the head is fixed in a definite way.

We have carried out an anthropometrical study of the skull in a male patient, aged 30 (Figures 1a and b). In this study the following requirements were fulfilled:

- the head was placed in close contact with the cassette holder;
- the head was fixed by ear moulds at the level of the external auditory canals and by an occipital head support;
- the head was positioned in such a way that the line between the external ear canals was parallel to the x rays;
- the distance between the cassette holder and x rays tube was 4.50 m, resulting in an optimal distortion of 3%;
- the patient had his teeth in centric occlusion.

RESULTS

The results of these measurements are given in Table 1.

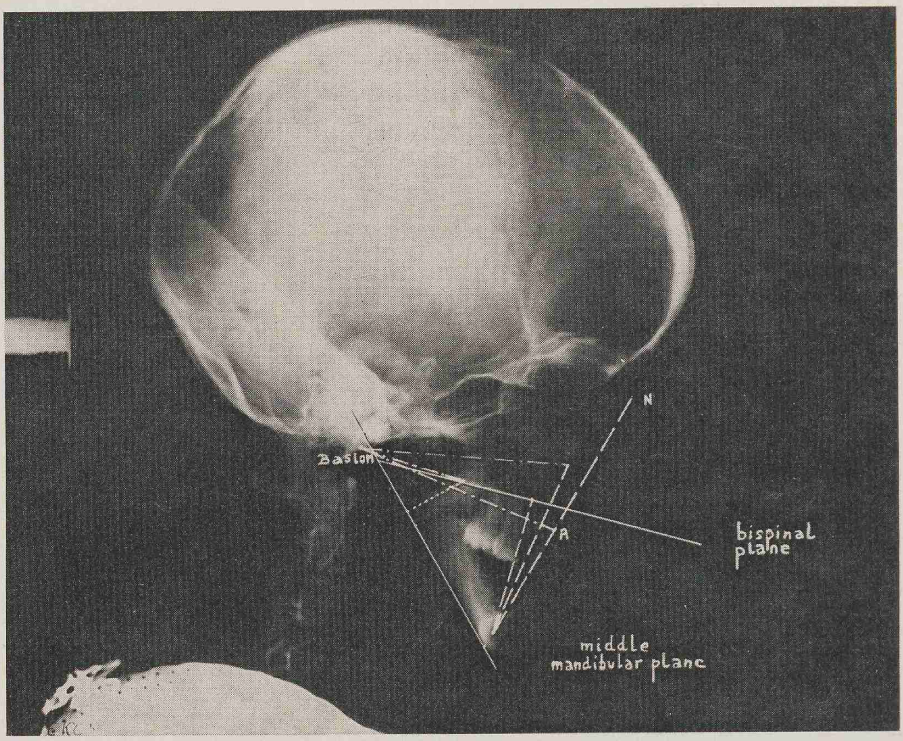

Figure 2.

Table 1.

measurements	patient	normal	% average norm.
Angular measurements (degree)			
angle of the base of the skull (Basion, pituit.tuberc., Nasion)	141°	132°	
angle SNA	68°	83°	
angle SNB	68°	80°	
angle ANB	0°	3°	
angle of Schwartz (Francf. plane, bispinal plane)	N	± 0°	
angle of Tweed Francf. plane, mandibul. plane)	45°	22°	
mandibular angle	146°	120°	
	158°		
Linear measurements (mm)			
Base of the skull			
Basion, pituit. Tuberculum	41	52	78,8
pituit. Tuberculum, Nasion	52	64	81,2
Basion - Nasion	89	106	83,9
Maxilla			
length of the base (post nasal spine, point A)	36	51	70,5
height bispinal plane – infer. orbit. point	13	25	52
height bispinal plane – nasion	39	54	72,2
Mandibula			
length of the ascending branch	37	66	56
length of the corpus mandibulae	40	79	50,6
total length	74	122	60,6
Facial height			
post. nasal spine - mandibul. plane	19	50	38
symphysis - bispinal plane	49	69	71
symphysis - infer. orbit. rim	62	94	65,9
symphysis - nasion	91	124	73,4
Measurements starting from the basion towards facial landmarks			
basion - post. nasal spine	34	46	73,9
basion - inf. orbit. rim	67	82	81,7
basion - point A	70	96	72,9

The linear and angular measurements have been compared with the same measurements performed in 109 adult men in which the anterior dental articulation and the profile were considered to be within normal limits.
This latter study has also been performed by the use of the same radiological equipment (cephalostat).

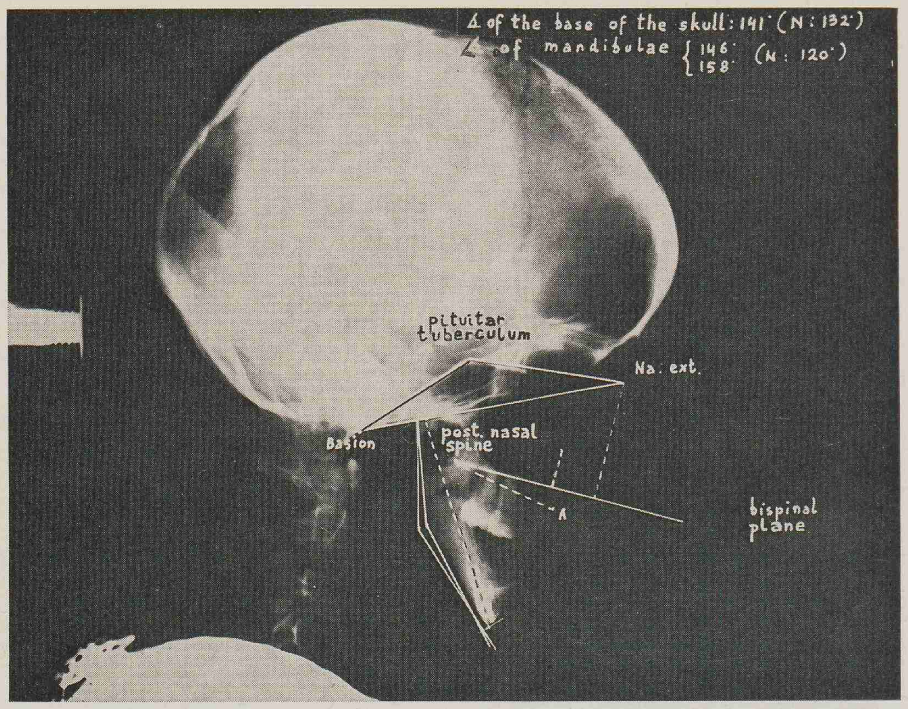

Figure 3.

Angular measurements (Figures 2 and 3): There exists a certain degree of platybasis (the angle of the base of the skull is increased). This angle, however, does not exceed the limits of the normal subjects in our study. The bispinal plane has normal angular relations with the Francfort plane and the Sella turcica - external nasion line. The most important anomalies are situated at the levels of the maxilla and mandibula. The localisation of the anterior maxillary limit is far behind the external nasion (angle SNA 68° instead of 83°) and the mandibula shows a very important retrognathia (angle SNB 68° instead of 80°). Furthermore, one can see a mandibular angle (146° and 158°) which is far superior to the normal average of 120°. This gives the mandibula a "spread out" spatial position.

Linear measurements (Figure 4): All measurements, without any exception, are reduced or much reduced in comparison with the normal averages. The base of the skull is the most normal skeletal zone. The most abnormal parts are the maxilla and the mandibula. As one encounters very often in case of micromandibulia, here too the posterior facial height (height of the ascending branch) is much more involved than the anterior facial height.

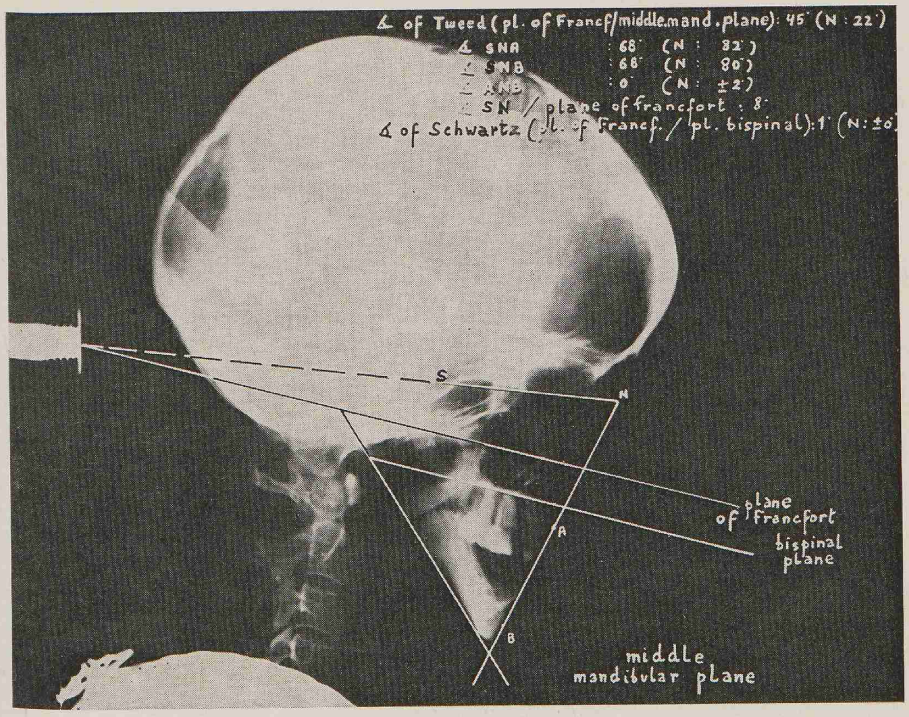

Figure 4.

DISCUSSION

Pathogenesis: From the literature follows that the syndrome results from an ecto-mesodermic hypoplasia occuring mainly between the 5th and 7th week of the embryological development, involving especially the first branchial arch (Hallermann, 1951; Ullrich and Fremery-Dohna 1955; François, 1958; Calmettes et al., 1960; Guyard et al., 1962).

Etiology: The etiology of the disease still remains obscure. There does not exist any preponderance as far as sex or consanguinity are concerned. Some authors have suggested a viral embryopathy, but this hypothesis has not been proved until now.

In this patient a karyotyping has been performed which turned out to be normal. The normality of the karyotyping, however, does not exclude the hereditary transmission of this desease.

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