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Hypertelorism, a hitherto  
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cranio-facial deformity**

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## HYPERTELORISM.\*

*A hitherto undifferentiated congenital cranio-facial deformity.*

By DAVID M. GREIG, C.M., F.R.C.S.E., Conservator of the Museum.†

*Any event in this world—any human being for that matter—that seems to wear even the faintest cast or warp of strangeness, is apt to leave a disproportionately sharp impression on one's senses.*—Walter de la Mare.

I DEEPLY appreciate the honour that has been conferred upon me in asking me to deliver the Struthers' lecture. Sir John Struthers was a life-long friend of my father, and as Professor Struthers he formed one of the earliest recollections of my childhood and remained thereafter a paradigm and an incentive. It was an admirable characteristic of Sir John Struthers' anatomical studies, a characteristic thoroughly appreciated by surgeons, that they so frequently presented a clinical vein. No one knew better than he that the study of anatomy necessarily entails the study of abnormalities, that an understanding of growth is interwoven with appreciation of congenital defects, that the so-called normal is but the average among variations, and that the abnormal is merely a departure from a somewhat arbitrarily chosen type.

I am not able to follow Professor Struthers' example by making the anatomical aspect the primary one, but I hope to present a clinical study in which I believe anatomical considerations do not form the least important feature.

The outstanding peculiarity of the cranial deformity for which I propose the name ocular hypertelorism, or briefly, hypertelorism, is the great breadth between the eyes. Hypertelorism ( $\acute{\upsilon}\pi\acute{\epsilon}\rho$  = too much,  $\tau\eta\lambda\epsilon$  = apart,  $\acute{o}\rho\acute{\iota}\xi\omega$  = to separate) preceded by the adjective ocular, accurately, concisely, and suggestively describes the salient and constant feature observable during life.

Great breadth of nasal bridge sometimes with hypertrophy of bones, interocular hyper-rhinoplasty, to coin another name, may exist without an undue distance between the eyes. The eyes may be far apart in cases of frontal meningocele or encephalocele, or in cases of congenital facial or nasal cleft,

\* The fourth Sir John Struthers' lecture delivered in the Hall of the Royal College of Surgeons of Edinburgh, 1st February 1924.

† I desire to thank the Carnegie Universities Trust for a grant towards the cost of reproducing the illustrations of this paper.

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but the ocular far-apartness is then subsidiary to the tumour or to the still more hideous defect.

CASE. I.—Mary Macdougall, who is the main subject of this lecture, was a native of Edinburgh, the only and illegitimate child of a feeble-minded mother. Her alleged father—a cabman—was addicted to occasional intemperance. Her maternal grandmother was not known to present any abnormality.

When about 7 years of age photographs of her (Figs. 1 and 2) were shown by Dr John Thomson, at a meeting of the Edinburgh Medico-Chirurgical Society (1) as those of an imbecile girl with a peculiar congenital malformation of the face. Dr Thomson considered that "the general look of the child's face recalled so strongly that of an early fœtus that it appeared likely that the whole thing was a mere arrest of normal development of some kind," while Dr J. W. Ballantyne "was inclined to regard it as a slight one of 'rhinodyme,' *i.e.*, an attempt at doubling of the parts at and above the level of the nose." (*Cf.* 39, 40.) I was much struck by the photographs, and very interested in the opinions then expressed, and a few years later when the child was transferred to my own care I received her with a re-awakened interest.

When she was 16 years of age she was undersized but not excessively so. Her skin, though soft and smooth, was of a dusky grey colour and her extremities presented an acrocyanosis of a pinky-blue tint. Her fingers and toes were short in comparison with their breadth and were unusually soft and fat. She had large hands and a fairly powerful grip. The subcutaneous tissue was well developed especially about the neck and chin. The distance between the eyes, and the great breadth of the lower part of her nasal bridge (Fig. 3) have been already referred to as the outstanding features of hypertelorism. The nasal bridge was flat and the nose in consequence *retroussé*. The vertical groove which Dr Thomson had noted as being so much in evidence nine years previously, and on which Dr Ballantyne, perhaps too greatly, had based his diagnosis of "rhinodyme" was not nearly so distinct. The forehead formed a regular anterior curve with slight prominence of the frontal eminences (Fig. 4). The eyelids were somewhat full and fatty and gave the impression that the eyeballs were overloaded by them. She had a bilateral external strabismus, the eyes being apparently equally affected (Fig. 5). Her eyes did not converge on near objects. Like a hare, she could not see objects directly in front of her so readily or so well as when they were placed laterally or when she turned her head away from them. The movement of the eyeballs did not always co-ordinate. I was never successful in getting a view of the fundus oculi, but believe there may have been some optic atrophy, as towards the end of her



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life her eyesight failed considerably. Her tongue was large and slightly fissured as in tongue-sucking children. Though Dr Thomson had noted her teeth as good and normal, they were very bad, carious, and defective when she came under my observation. She never complained of toothache but the teeth loosened and many disappeared. Puberty came on about 15 years of age and her breasts became loose, pendulous, and flabby. Menstruation gave her no trouble. She was twice operated on for tuberculous cervical adenitis. She died at the age of 22 years from pulmonary tuberculosis.

To many casual observers her countenance was repellent, but to those in daily contact with her her gentle and affectionate nature made her an acceptable child. Always even-tempered, she could be trusted with even some of the most restless children in the Institution, and these she used to exercise in the grounds and she seemed quietly alive to her responsibilities when her charge thought fit from time to time to break away. She had cleanly and tidy habits, was fond of a pretty dress or coloured ribbon, and was modest and of a retiring disposition. She used to help to dress and undress the helpless children, and could nurse and feed them until her eyesight became too deficient.

A little knitting was the only handiwork in which she could take an interest, and of this she accomplished very little. Her hearing was good and she enjoyed music of any kind. Her vocabulary was limited and her articulation very defective, at best being little more than a mutter. Her voice was rather low-pitched. Her memory was fair. She could give the names of some of the children and nurses with whom she was associated, but questions when answered at all were replied to in single words only.

In this individual there is a congenital cranio-facial deformity with mental deficiency, associated as is so often the case with general retardation of growth, peculiarities of the extremities and with autonomic vascular disturbances as evidenced by the acrocyanosis.

A case of the same deformity but of rather less degree is the following:—

CASE II.—Annie C. (Fig. 6) was 10 years old when brought to me on account of tuberculous cervical adenitis, which had followed an operation for otitis media four years previously, but the notes of her deformity were taken when she was 12 years of age. She was the third of eight children. A younger brother died of pneumonia, aged 4 months, and the youngest of the family, aged 6 weeks, was said to take right unilateral convulsions. These children and the parents were all well formed. Annie was of fair stature, height 4 ft. 7 in., weight 5 st., active and lively, and took an intelligent interest in her surroundings. She had a somewhat narrow forehead,

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with slight prominence of the frontal eminences which towards the root of the nose were separated by a shallow groove. The hairy scalp formed a peak in the middle line which encroached on the forehead. The eyebrows were of unusual vertical depth. She had a habit, apparently involuntary, of raising the eyebrows spasmodically together or independently of each other, but this happened much more often and to a greater degree with the left than with the right, and may have had its origin in attempts to overcome the ptosis. As is seen in the photograph this was accompanied by corrugation of the skin over the corresponding belly of the occipito-frontalis. The eyes were far apart, the nasal bridge low and broad, and there was a shallow vertical groove or perhaps merely a flattening along the nose (Fig. 7). The narrowness of the forehead gave the orbital contents the appearance of bulging laterally. The upper eyelids were large and full, and of the left the folds were less distinct and there was a slight ptosis. The right palpebral slit was somewhat oblique. The pupils were dilated, the right rather larger than the left, and both reacted slowly and imperfectly to light. The inner canthi were 50 mm. apart and the palpebral slit measured 40 mm. The right eye had a tendency to lateral deviation, while from deficient power of the lateral rectus muscle lateral deviation of the left globe was restricted. The occiput was somewhat flattened and the circumference of the head was 510 mm. She kept her mouth slightly open and her lower lip and mandible projected (Fig. 8). Her sight and hearing were good, and her teeth normal and well formed. She answered questions readily but talked in a somewhat childish manner and as if her tongue were too big, as indeed it appeared to be. Her palate was perhaps slightly more arched than normal. The little fingers were disproportionately small, and all the digits ended with unusual squareness especially the thumbs. She was in the third standard at school but could not tell her age. She said she had never learned to sew but that she could knit. Operation on the glands of the neck was followed by tuberculous axillary adenitis which was in turn operated on. Death resulted from tuberculous cervical spinal caries with diplegia at the age of 17.

Of these two examples of hypertelorism it is apparent that that of Mary Macdougall is the more exaggerated. That they are different degrees of the same deformity there is no reason to doubt, and the radiograms of the skull taken during life further support this assertion. In the more advanced condition (Fig. 9) the forehead is lower, the occiput is flatter, but altogether there is a striking similarity between the two (Fig. 10). The radiograms give the impression that the skull is of more than average thickness, but, as will be seen



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presently, examination of the macerated skull does not bear this out.

Scattered here and there throughout medical literature are illustrations of children who seem to present the characteristic deformity of hypertelorism but the cases are described as examples of "oxycephaly." For example, the photograph, and to some extent the description of Case I. in Hutchison's paper,<sup>35</sup> is much more suggestive of hypertelorism than of oxycephaly. The child, a female, aged 1 year, was of doubtful mentality. Park and Powers<sup>2</sup> write: "In the photographs which accompany the case-reports the eyes appear to be far apart," but the *reports* do not always state that. The impression conveyed by a photograph is not entirely trustworthy, and an appearance of far-apartness may be produced by an external strabismus (a common accompaniment of oxycephaly) and by flatness of the nasal bridge. The converse also holds good, for in a case reported by Young<sup>3</sup> the eyes are said to be far apart, yet the accompanying photograph scarcely bears that out. Not that these cases are all advanced as typical cases of oxycephaly, but they evidently seemed to their chroniclers to be more in consonance with that deformity than with any other recognised cranio-facial defect. Neither in oxycephaly nor in rhinoplasty is far-apartness of the eyes a necessary attribute, in hypertelorism it is essential. Nevertheless these cases and such weight of authority led me for a time to consider this deformity as an atypical variation of oxycephaly, and it was not until I had the opportunity of studying Mary Macdougall's macerated skull that I realised I was in the presence of something entirely different.

As I pointed out in a demonstration to the Medico-Chirurgical Society of Edinburgh, on 7th December 1921, oxycephaly is commonly made to include two totally different conditions. In true congenital oxycephaly synostosis takes place at all the skull sutures—the facial as well as the cranial—as soon as the growth of the bones brings them into contact with each other, be they developed in cartilage or in membrane. Under ordinary conditions of growth as the bones approximate they form sutures which are present until full growth is reached, but in true congenital oxycephaly where the bones touch they fuse. The growing brain may and does keep the bones, especially the membrane bones, apart as long as it can, for life depends on the brain finding room to expand. The term "premature



FIGS. 1 and 2.—Hypertelorism ; Case I., aet. 7 years.  
 FIGS. 3 and 4.—Hypertelorism ; Case I., aet. 18 years.

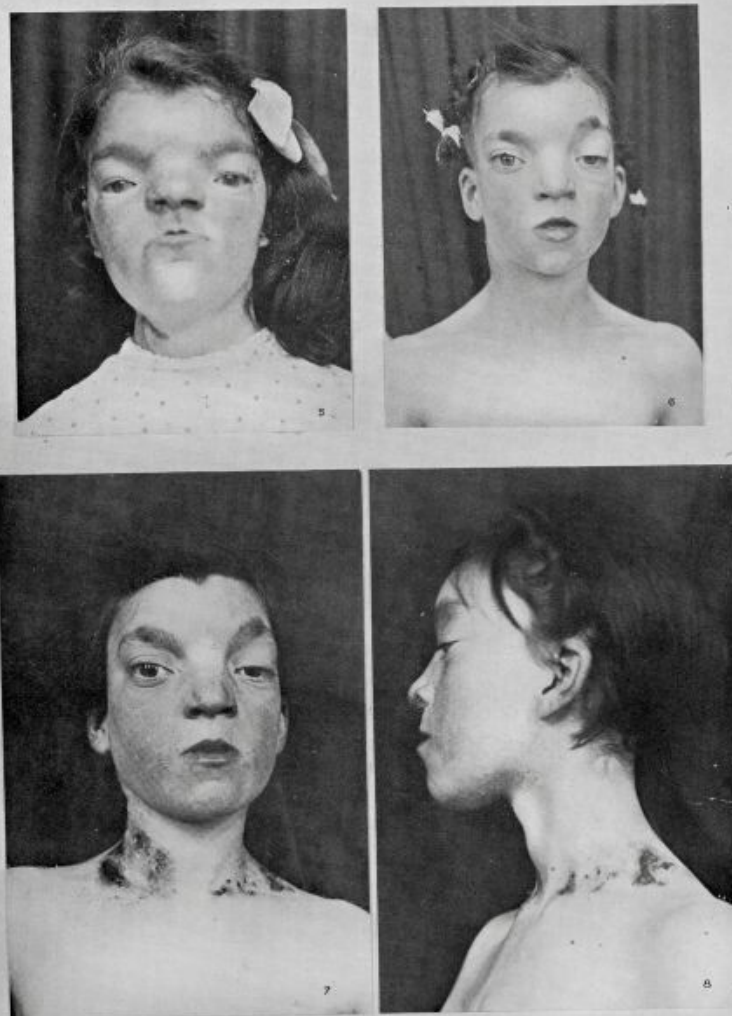


FIG. 5.—Hypertelorism; Case I., æt. 16 years.  
 FIG. 6.—Hypertelorism; Case II., æt. 12 years.  
 FIGS. 7 and 8.—Hypertelorism; Case II., æt. 15 years.



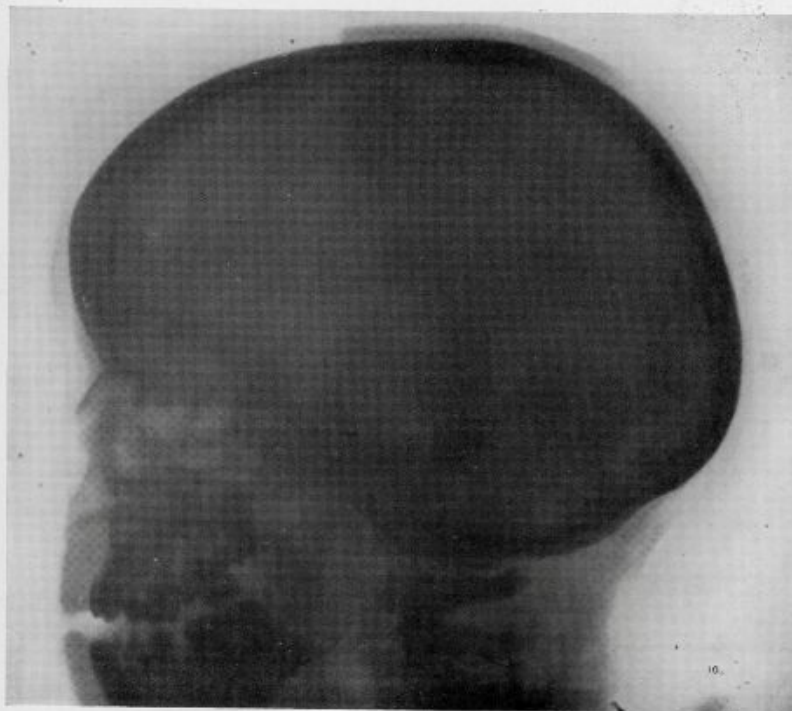
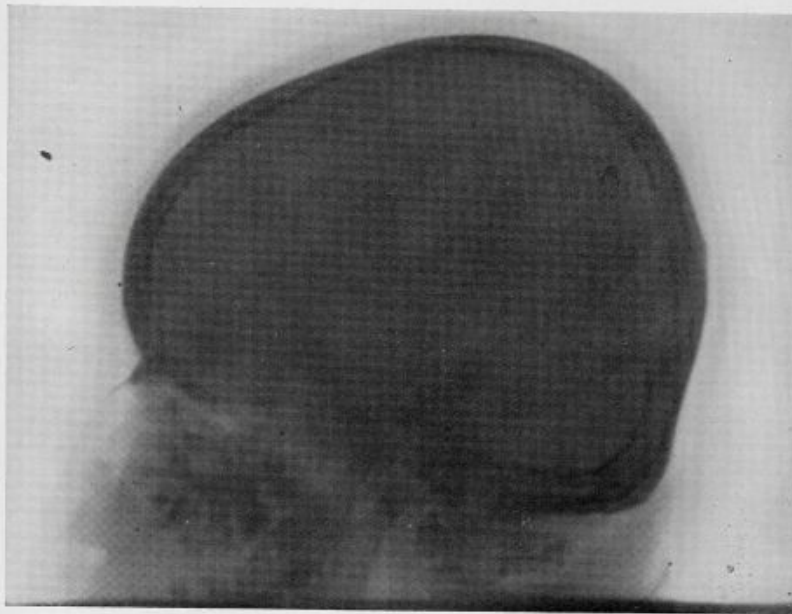


FIG. 9.—Hypertelorism ; radiogram of Case I. during life.  
FIG. 10.—Hypertelorism ; radiogram of Case II.

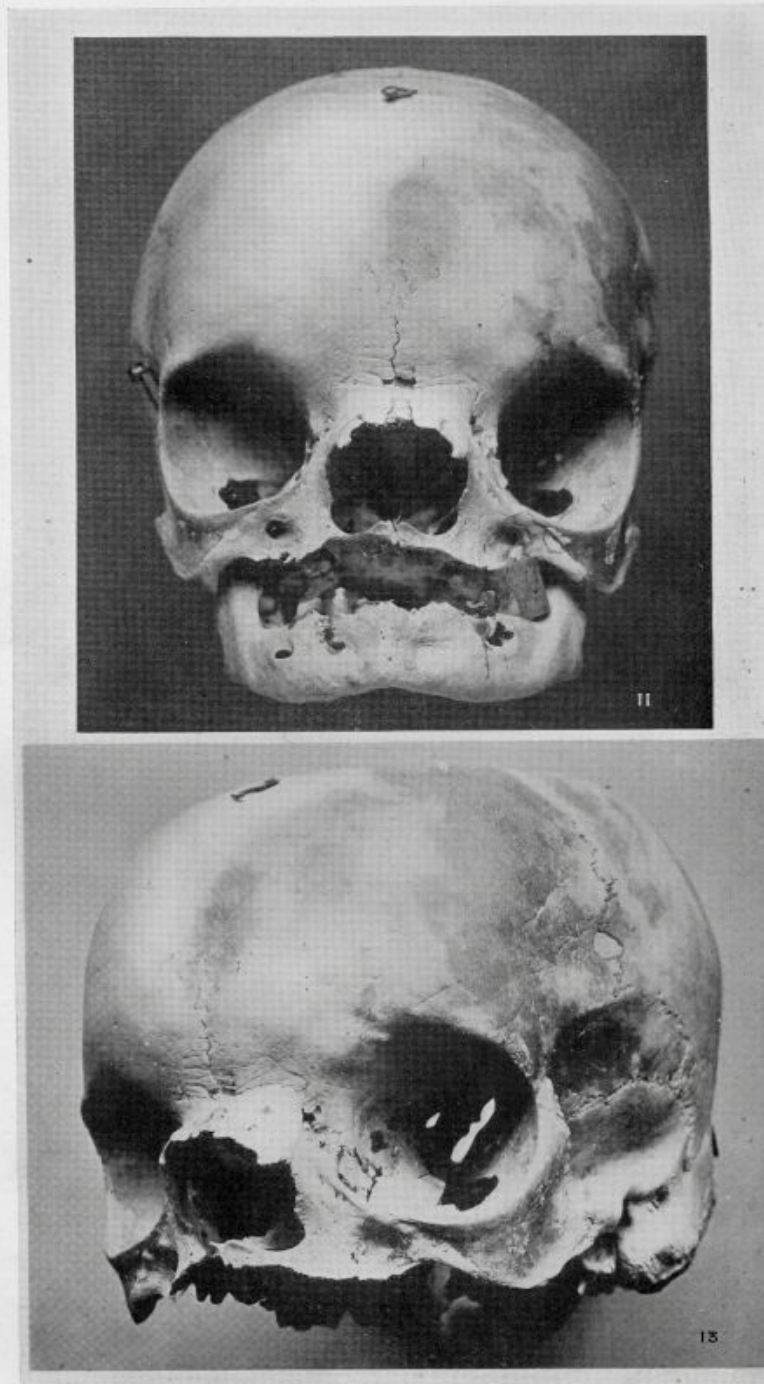


FIG. 11.—Hypertelorism ; norma frontalis  
 FIG. 13.—Hypertelorism ; left orbit.

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synostosis" inadequately expresses the condition in true oxycephaly. Sutures never really exist as such. Wherever possible the bones are pushed apart by the growing brain, wherever possible the bones yield to pressure and the fontanelles and inter-osseous spaces are widened. Across these spaces bony fibres extend, but when opposing fibres meet they do not interlock but become incorporated to form one continuous bone. It follows that the characteristic deformation begins at the skull-base and the shape of the skull is modified along the lines of possible cerebral expansion.

The other form is a pseudo-oxycephaly. It confuses the appreciation of true oxycephaly, for in it synostosis is limited to one or more sutures and occurs later in life though before the brain has attained its maximum growth. Pseudo-oxycephaly is never a general affection of the skull sutures, it is unassociated with defects of the extremities, the base of the skull is not altered, and the facial bones are never involved. Here certainly we have a "premature synostosis," for the sutures have at one time been present. This deformity is a caricature of true oxycephaly. The term "premature synostosis" might, with advantage, be substituted for "pseudo-oxycephaly," for it would then have a wider application and ambiguity be avoided. I repeat that in true oxycephaly the characteristic is want of formation of sutures throughout the *whole* of the skull, exclusive of the mandible. It would not be expected that the mandible, whose evolution is so different from that of the skull and facial bones, need participate in their extravagances.

A mere glance at Mary Macdougall's skull is sufficient to show that we have here no oxycephaly for all the sutures are present, they are well formed and the deformation of the cranial cavity in no way corresponds to that of oxycephaly.

But the skull presents as a whole and in detail many interesting peculiarities.

### The Skull.

In the macerated skull the absence of eyeballs renders the hypertelorism less obvious, while the absence of nasal tissues and cartilages makes the unusual size of the nares and the breadth of the nasal bones obtrusive (Fig. 11). In addition to the peculiarities relative to hypertelorism the skull naturally presents subsidiary peculiarities such as may be found in



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many "normal" skulls, and these fall to be discounted. The characteristic features of hypertelorism are interdependent and form a definite anatomical combination.

The skull presents on inspection certain general features which are seen at a glance, namely, a low forehead, a pronounced



FIG 12.—Millimetre-scale drawing of the left lateral aspect of the skull in hypertelorism. The lines indicate distances of 5 cm.

vertex rising more posterior to the bregma than usual, and a flat occiput (Fig. 12). Craniometry also indicates a departure from the normal.

The maximum horizontal circumference is 352 mm., measuring round the most prominent parts of the glabella and occiput. The greatest length is 151 mm., measuring again from the most prominent part of the glabella to the most prominent point at the occiput, and the greatest parietal breadth, which

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is behind and above the temporal line, is 124 mm. These measurements give a cephalic or longitudinal index of 82, and the skull is therefore brachycephalic.

The biasterionic width is 103 mm. and the bistephanic is 119 mm. The maximum height of the skull is 122 mm. and the vertical or altitudinal index is 80. The skull would be classified as acrocephalic to use Turner's nomenclature; or hypsocephalic to use the term employed by Virchow and others, this latter term being surely derivatively preferable.

The basinasal length is 96 mm. measuring from nasion to nasion and the basi-alveolar length is about 87 mm., accuracy being unattainable as absorption of the maxillary alveolar processes has obliterated the true alveolar point. This measurement has consequently been taken from the nasion to the alveolar point. If then this measurement be utilised, the alveolar or gnathic index is 90 and the skull orthognathous.

The maximum bizygomatic width is 109 mm. and the total facial length from the nasion to the pogonion is 61 mm. or from the ophryon to the pogonion 70 mm., which makes the total facial index 55 or 64 according to which length-measurement of the face be adopted. The absence of alveolar point makes it impossible to obtain the superior facial index.

**Norma Frontalis** (Fig. 11).—The lowness of the rather narrow forehead is somewhat obscured by the prominence of the frontal eminences, and an appearance of breadth is imparted to this portion of the skull. Immediately above the root of the nasal bones is an opening 7 mm. in transverse and 3 mm. in vertical diameter. There is no frontal spine. Though freely communicating with the interior of the skull by its upper half, the lower half of this opening exposes the anterior aspect of the alar processes of the ethmoid. It takes the place of the foramen cæcum and may have merely contained a process of the falx cerebri and the initial vein of the superior longitudinal sinus. On the other hand it may be in relation to the anterior neuropore, for Rauber (quoted by Schultz<sup>4</sup>) believes that the anterior neuropore of the medullary canal of vertebrates can exert an influence under special circumstances even to the ossification of the skull. Its margins are irregular, and from a little to the right of the mid-point of its upper border a metopic suture passes upwards for 17 mm. The suture does not form digitations, the two halves of the frontal being merely approximated and only at its upper end do the bones come

actually into contact. The metopic suture terminates above in a triradiate divergence, one limb passing directly upwards while the other two incline obliquely on either side. The limbs each measure about 8 mm. in length and may indicate the remains of a metopic fontanelle. The contour of the glabella forms a regular anterior convexity and in this part there is no mesial groove. The prominence of the frontal eminences, however, gives the appearance of a broad and shallow sulcus in the upper part of the pars frontalis, but the sulcus exists only in relation to the frontal eminences and is not continued to the bregma.

(a) *The Orbits*.—Making allowance for the fossa saccularis lacrimalis the margin of each orbit is almost circular. In three quarters of its circumference the edge is much more sharp than usual, while in its upper and mesial segment it is rounded and slightly undulating. The supra-orbital notch is a shallow crescent in the left (Fig. 13) and scarcely discernible in the right orbit (Fig. 14). It is situated 40 mm. from the sagittal plane, a great distance in such a small skull, since according to Schwegel (quoted by Whitnall<sup>5</sup>) 40 mm. is the maximum in an adult skull of average growth. There is no trace of orbital tubercle; perhaps the lateral deviation of the eyes and orbits did not call for its development. A short shallow groove on the left and two fainter and more shallow grooves on the right lateral aspect of the frontal pass backwards and laterally for the accommodation of branches of the supra-orbital nerves.<sup>6</sup> The posterior margin of the fronto-sphenoidal process of the zygomatic is serrated. The zygomatic canal is present on the right side but absent from the left. The lateral margin of the left orbit lies 32 mm. and of the right 24 mm. behind the coronal plane at the root of the nose. This indicates a lateral deviation of the orbital aditus far beyond the normal.

On the roof of the orbits the fossæ for the lacrimal glands have not retained the conical shape found in the foetus. At the apex of the left fossa is a minute foramen, and at the apex of the right fossa a slit divided by a small spicule of bone into two small foramina. The superior of these two small foramina passes upwards and laterally into the cranial cavity and finds its exit 1 mm. lateral to the lateral extremity of the small wing of the sphenoid, and there it becomes continuous with a branched venous groove which ascends a little way before turning forwards to the region of the frontal eminence. The inferior of these foramina in the right orbit passes more



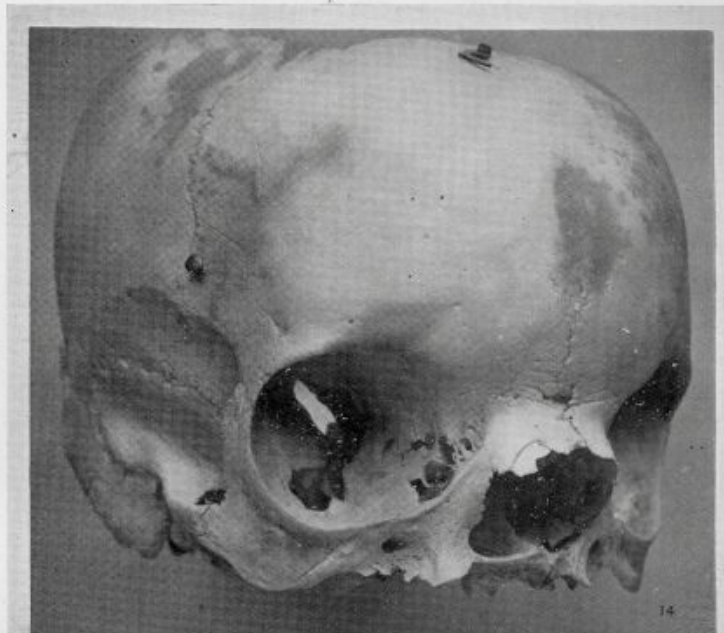


FIG. 14.—Hypertelorism ; right orbit.

FIG. 15.—Hypertelorism showing vertical division of left lamina papyracea.

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directly backwards and debouches below the level of the posterior free border of the small wing of the sphenoid into the sphenoparietal sinus. The small foramen in the lacrimal fossa of the left orbit corresponds to the superior of the two just described, and entering the cranial cavity passes a little way in a tunnel under the internal table and ultimately forms a venous groove which is symmetrical in course and distribution with that of the right side. These foramina are "normal" variations, and in the normal skull of a boy aged 14 years I have an absolute replica of them as just described.

The fovea trochlearis is well developed in each orbit as a smooth plain depression. The posterior limb of the fronto-sphenoidal suture is still present. The optic foramen is pyriform with its apex directed upwards. The roof of each orbit is exceedingly thin and contains no sinus-extension. Though, as is usual with transmitted light, the juga cerebralia and the cerebral impressions between them are clearly seen, the translucency is not confined to the posterior two-thirds but extends over the whole roof. On the right side the zygomatic canal passes from the orbit obliquely forwards but there does not seem to be a posterior division, while on the left side it is unusually small and consists of a posterior division only which passes almost directly backwards to open into the temporal fossa deep to the temporal process of the zygomatic bone. There is no anterior division, nor is there on the surface a zygomatico-facial foramen. In each orbit a meningeal foramen is situated in the fronto-sphenoidal suture a little way in front of the anterior termination of the superior orbital fissure.

Though this is the skull of a young person, both the lateral wall of the orbit and the floor are materially encroached upon by the large inferior orbital fissure. This allows of unusually free communication between the orbit and the temporal fossa, as is present in the foetus and in so many of the lower mammals.

Each superior orbital fissure is 17 mm. in length and more simple than usual. Its upper and lower borders lie parallel, and it terminates laterally and mesially in smooth concavities of equal size. The anterior opening of the foramen rotundum is easily seen on looking into the orbit from the front, an indication that the bony parts are more spread out than usual.

On looking into the orbit two foramina, larger in the left than in the right, can be seen on the posterior root of the small wing of the sphenoid lateral to the optic foramen. In the left

orbit the lower of these foramina opens within the cranial cavity 3 mm. behind the centre, and the upper opens 5 mm. lateral to the apex of the pyriform optic foramen. In the right orbit the lower foramen does not enter the cranial cavity but ends in the cancellous tissue of the sphenoid, while the upper foramen has the same course as its counterpart in the left orbit. These foramina indicate additional venous channels. There is no zygomatico-orbital foramen.

An infra-orbital suture is present in each orbit, but in the right it only extends a little way from its commencement at the inferior orbital fissure, while in the left it can be traced over the orbital margin into the intra-orbital foramen.

On the mesial wall of the orbit the fronto-ethmoidal suture is interrupted about its centre by a large quadrilateral intersutural bone which extends upwards on to the roof. It measures in its vertical diameter 15 mm. and in its horizontal diameter 12 mm. It is a single bone in the right orbit, but in the left orbit (Fig. 15) is halved by a vertical suture, but apart from this division its symmetry in the two orbits is very striking. According to Whitnall,<sup>7</sup> vertical division of the lamina papyracea was "found by Bianchi (quoted by Le Double) to occur more frequently in the skulls of imbeciles,"<sup>37</sup> and this I can confirm from my own observation, but the quadrilateral bones just described are not of this nature, they are true intersutural bones and not divisions of the ossa plana. The anterior part of the lamina papyracea is exceedingly delicate, and in many places so deficient as to give it a fenestrated appearance. The lacrimal bones are small and light and are fenestrated, which, according to Macalister, is normal in the foetus.<sup>8</sup> The anterior and posterior ethmoidal canals cannot be identified in either orbit, but apparently are incorporated with the fronto-ethmoidal suture, for their intra-cranial openings can be recognised though they are very small. As these canals are so constantly present, and as they transmit definite nerves, their absence would be a matter for surprise.

The orbits are of considerable depth, measuring to the orbital edge of the optic foramen in the left orbit 38 mm. and in the right 40 mm. (Fig. 16). The orbital height is 37 mm. and the greatest width 38 mm. The orbital index is thus 97, and the orbits are therefore classed as megaseme.

(b) *The Nasal Cavity*.—The nasal bones are short, broader than they are long, and the whole internasal suture is occupied



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by a narrow intersutural bone which lies almost horizontally (Fig. 17). Each of the nasal bones measures in its greatest diameter 13 mm. sagittally and 15 mm. coronally, and the internasal bone is 7 mm. long by 5 mm. broad. The internasal bone is prevented from articulating with the frontal by a spur

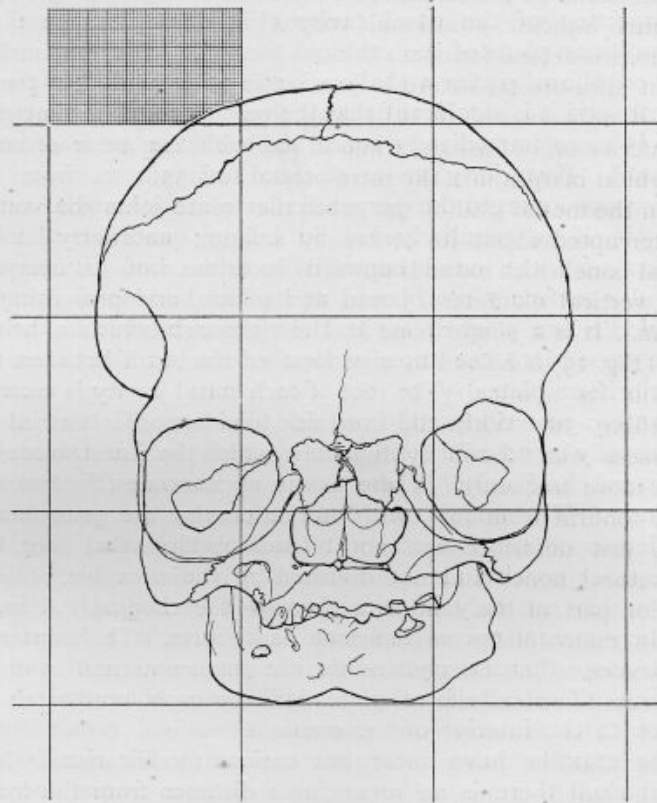


FIG. 16.—Millimetre-scale drawing of the norma frontalis of the skull in hypertelorism.

which passes mesially from the proximal end of the mesial border of each nasal bone. These spurs come very close to each other but do not interlock. The bony nasal bridge gives no indication of a mesial groove.

The enormous breadth of the anterior nares is the most striking feature of the skull. The name "apertura pyriformis" is, in this instance, entirely unsuitable, for the opening is quadrilateral. Absence of a septum nasi from the anterior part

and absence of the maxillary alveolar processes accentuate the size of the nasal aperture. The nasal height, measured from the nasion to the lower margin of the nasal opening, is 30 mm. and the greatest width is also 30 mm. The nasal index is, therefore, 100, and as anything above 53 constitutes "platyrrhine," this skull is hyperplatyrrhine to a remarkable degree (Fig. 16).

Inspection of the nasal cavity (Fig. 18) shows that the perpendicular plate of the ethmoid forms on the under surface of the cribriform plates a shallow keel not exceeding 2.5 mm. in depth, and it is significant that the part retained is that part which is more immediately concerned with the sense of smell in man.<sup>9</sup> Posteriorly at a distance of 33 mm. from the anterior nasal margin the perpendicular plate joins the vomer, which itself is peculiarly low in height; anteriorly at its junction with the nasal bones it broadens into an irregular triangular surface 7 mm. broad at its base, and presenting in the middle line a small sagittal slit through which a bristle can be passed into the anterior fossa of the skull between the two cribriform plates. The roof of each nasal cavity is convex, both antero-posteriorly and from side to side, and is formed by an unusually broad cribriform plate in which the fenestræ occupy the lateral half only. There is no appearance of ethmoidal labyrinths. The middle and inferior conchæ are present and well developed, the latter almost touching the nasal floor, but the superior conchæ are represented by a diminutive, oblique ridge which can only be seen through the choanæ. A small maxillary antrum opens into each nasal fossa. The septum is well developed posteriorly, and the choanæ are normal in shape and size. The cribriform plates will be considered again in relation to the interior of the cranium.

The maxillæ have undergone curious modifications. In a normal skull there is an appreciable distance from the lower border of the zygomatic bone and the upper alveolar margin, and even in edentulous skulls there is some distance between these two points. But in this skull a straight-edge can be laid between the lower borders of the two zygomatic bones at any part and will not touch the palate. The skull deprived of the mandible and laid on a flat surface (Fig. 21) rests on the occipital condyles, on the tips of the pterygoid processes, and on the lower borders of the zygomatic bones, and neither the palate in front nor the occipital behind touches the surface on which the skull is resting. This is due to absorption of the

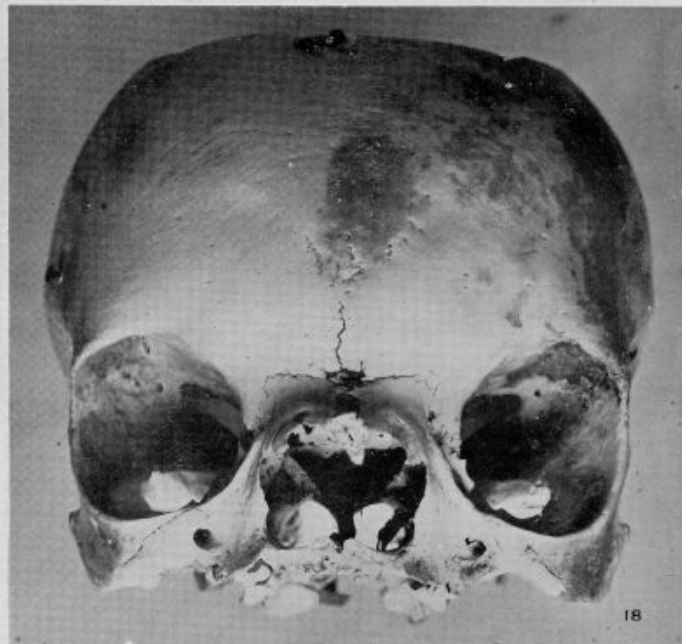
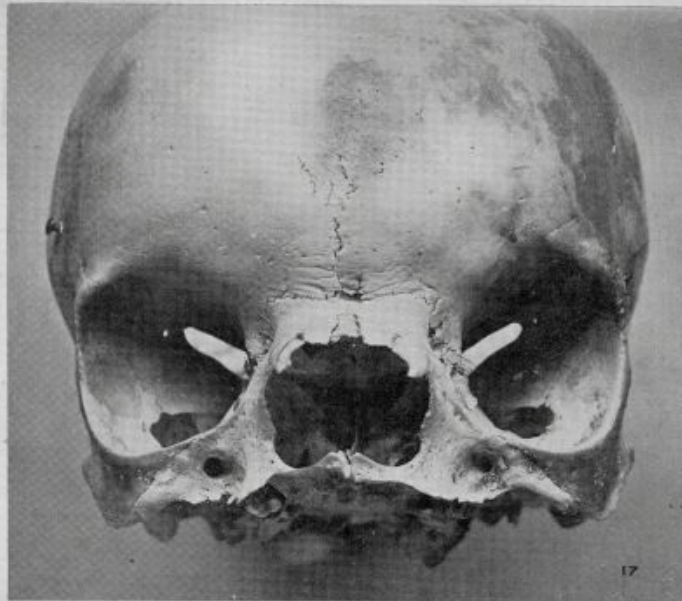


FIG. 17.—Hypertelorism showing the nasal and internasal bones and the size and shape of the anterior nares.

FIG. 18.—Hypertelorism showing the choanæ and the convex nasal roof.



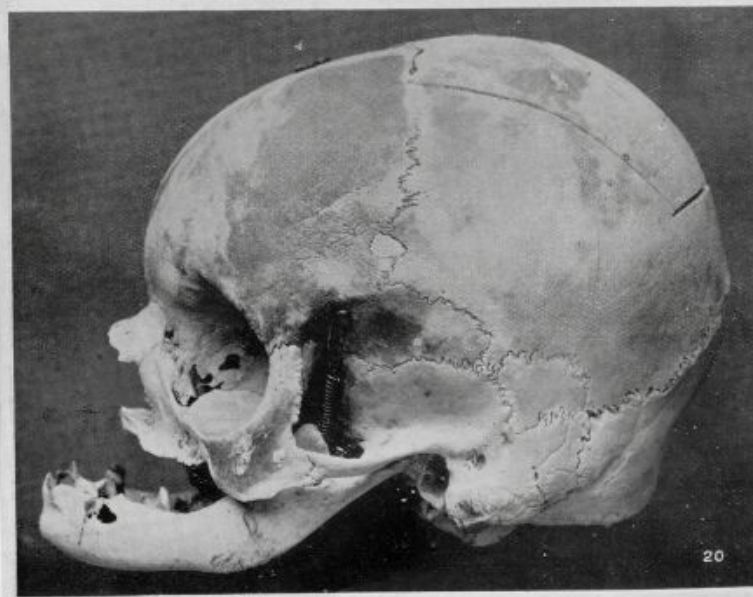
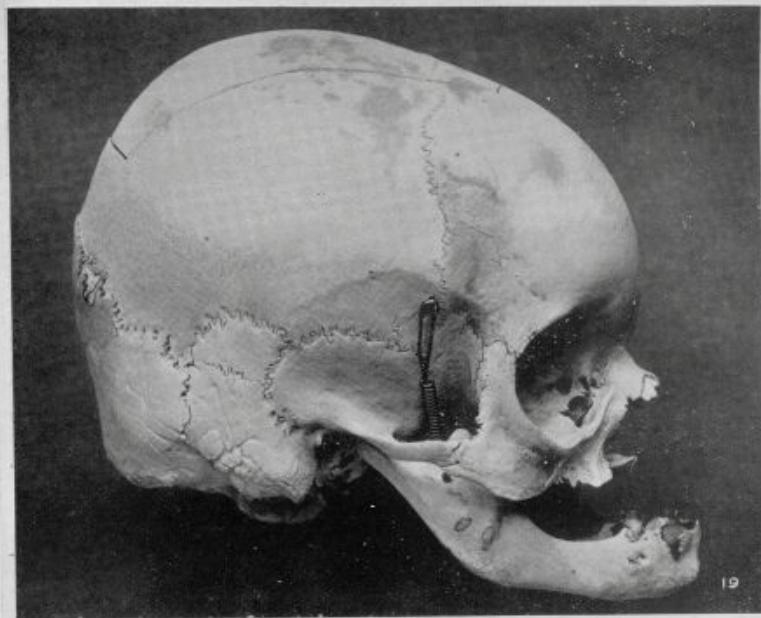


FIG. 19.—The right, and Fig. 20, the left, norma lateralis in hypertelorism.

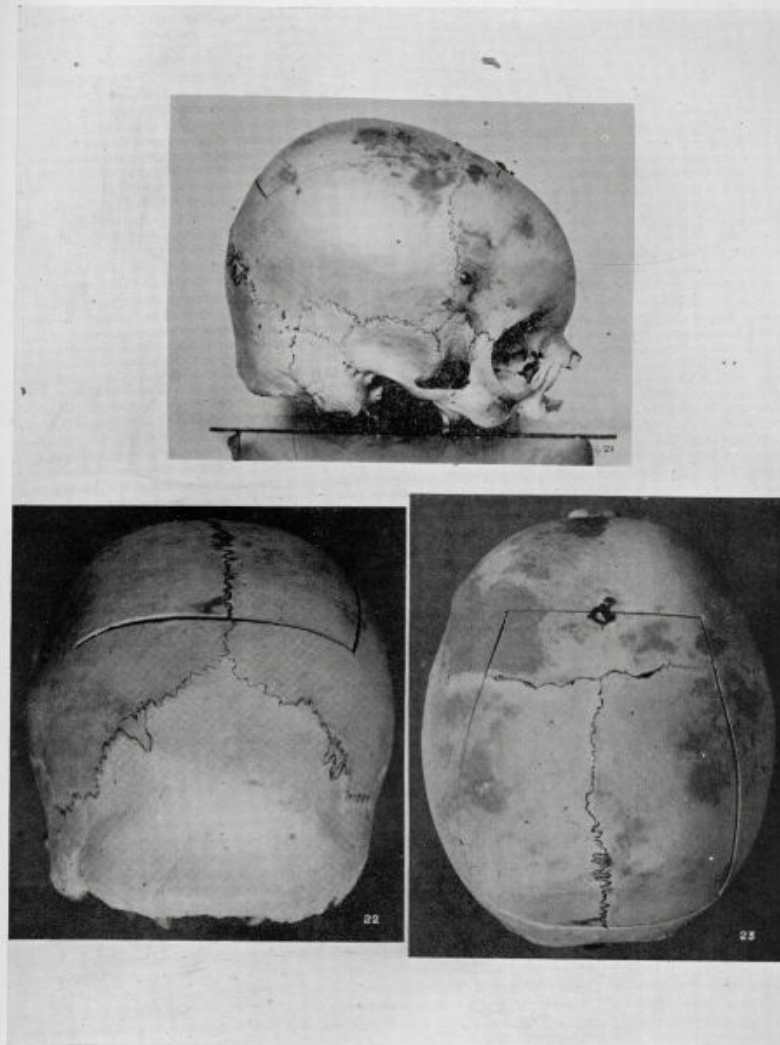


FIG. 21.—Hypertelorism ; the skull on a plane surface resting on its occipital condyles and zygomæ.  
 FIG. 22.—Hypertelorism ; norma occipitalis.  
 FIG. 23.—Hypertelorism ; norma verticalis.

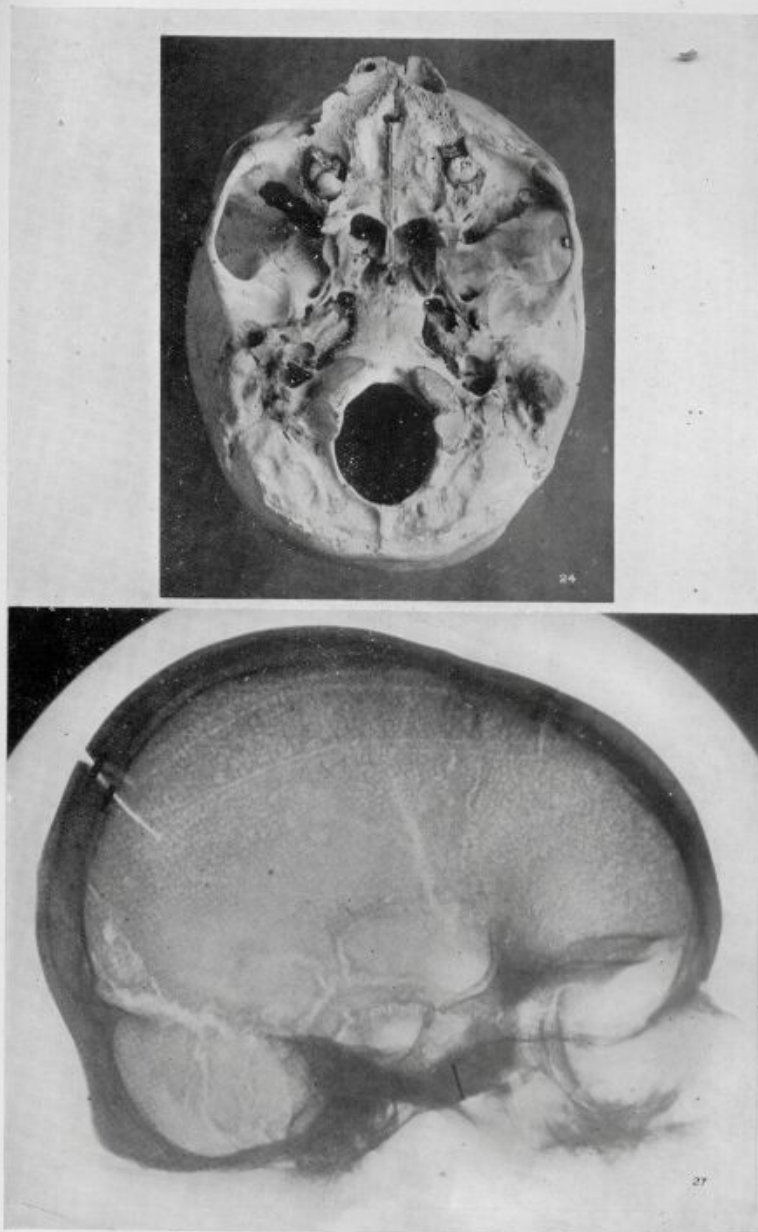


FIG. 24.—Hypertelorism ; norma basalis.

FIG. 27.—Radiogram of skull in hypertelorism. A metal rod in the cranio-pharyngeal canal is indicated by a black line in the skull base.



## Hypertelorism

alveolar processes to such an extent that their site is occupied by a concave broad sulcus of rarefied bone (Fig. 24). It is surprising to find any tooth in such a deterioration, but on each side a third molar is present, the left being carious and the right unerupted, and in front of each is the remains of a decayed second molar tooth. To such an extent has the alveolar destruction proceeded that at the nasal margin the left maxilla measures vertically only 2.5 mm., while the right is reduced to a linear edge. The anterior nasal spine is present and well formed, and as is so usual, the two halves slightly diverge from each other. The disappearance of so much of the maxillary body makes the abnormally large infra-orbital foramina look even larger. On the right side the opening is trumpet-shaped, and the canal so straight that one can look right along it into the inferior orbital fissure. The mesial wall or the left infra-orbital canal obstructs such a view, it is exceedingly thin and a small triangular area of it is deficient, so that the canal communicates with the maxillary antrum, though during life fascia and mucous membrane would have filled the hiatus.

**Norma lateralis** (Figs. 19 and 20).—The mastoid processes are comparatively massive, and each parieto-mastoid suture is entirely occupied by a large intersutural bone. The temporal line, which is pronounced on the frontal and anterior part of the parietals, has a low arc and ends at the anterior extremity of the parieto-mastoid intersutural bone. There is no mastoid foramen present on the left side, but on the right two small mastoid foramina lie in the centre of the occipito-mastoid suture somewhat hidden by the overlapping bone and open by a single opening into the sigmoid groove within the skull. The right external acoustic meatus is smaller and has thicker walls than the left. On the right side there is a small somewhat circular epipteric bone measuring 14 mm. in its greatest diameter. At the left pterion there is a large epipteric bone which extends up the coronal suture for 24 mm. Its upper border corresponds to the temporal line, and its greatest diameter, which is that passing antero-posteriorly, is 37 mm. In its backward projection it so nearly approaches the parieto-mastoid intersutural bone that there only intervenes a narrow tongue of parietal 5 mm. in breadth.

**Norma occipitalis** (Fig. 22).—The supra-inial portion of the occipital occupies nearly the whole field in this aspect of the skull and rises almost vertically from the lower part of the

squamous portion. The lambda is unusually high, and, except for a single symmetrical interposition at the junction of the supra-occipital with the inter-parietal portion, is devoid of intersutural bones. The inter-parietal portion bulges backwards beyond the supra-occipital portion, and of a slightly darker colour it appears less dense in texture, characters distinctive of its membranous development in comparison with the cartilaginous origin of the supra-occipital.

**Norma verticalis** (Fig. 23).—The skull is cryptozygous; its highest part lies behind the coronal mesial plane. The sagittal and coronal sutures present few denticulations, a not uncommon feature in the skulls of the congenitally mentally deficient. There are no parietal foramina, but dotted along the vertex a little way on each side of the sagittal suture are a few pin-point foramina which lead directly through the skull to the groove for the sagittal sinus or to the other venous channels on the intra-cranial surface. The very slight projection forwards of the right frontal region and backwards of the left occipital is such as is generally found in right-handed persons, and is the recognised normal asymmetry of the cranium. Where the cranium has been opened by raising a part of the calvarium the skull is seen to be of normal thickness, both tables distinct and the diploë normally developed and of normal consistency.

**Norma basalis** (Fig. 24).—The size and substantial development of the basilar process makes it appear disproportionately large. It measures 29 mm. in length and is 21 mm. in breadth just before it begins to widen towards the ex-occipitals (Fig. 25). The spheno-occipital synchondrosis is nowhere visible though a little irregularity of the bone indicates its position. The centre of the external opening of the cranio-pharyngeal canal lies 7.5 mm. in front of the line of union. In shape the opening is pyriform, with the apex directed forwards, and it measures 3 mm. at its widest part. The canal admits a wire 1 mm. in diameter and is 10 mm. in length. It is directed vertically upwards and opens a little in front of the deepest part of the pituitary fossa (Fig. 27).

The inferior wall of the left acoustic meatus is pierced just lateral to the site of the membrana tympani by a circular opening 4 mm. in diameter, and a similar though slightly smaller opening is present on the corresponding part of the right osseous meatus. This is the foramen of Huschke which normally disappears about the third or at latest about the fifth

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year of life. The anterior wall of the left meatus is thin and fenestrated.

The bone forming the floor of the carotid canal is light, diaphanous, and translucent on both sides, though this is less noticeable in the posterior half of the left canal. In quite half its extent the floor is deficient towards the foramen lacerum where the canal is replaced by a groove on the under surface of the petrous bone. The right jugular fossa is larger than the

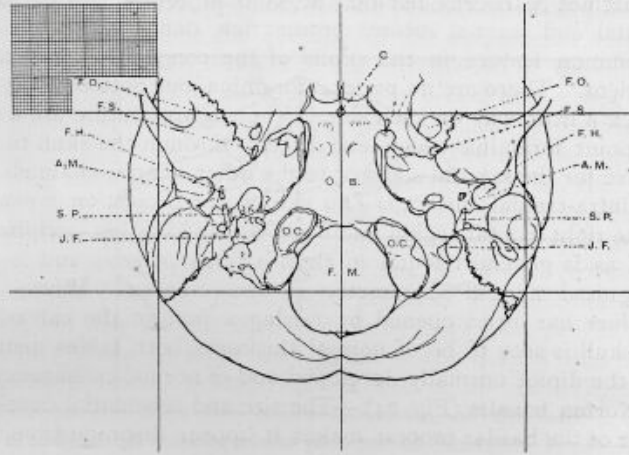


FIG. 25 — Millimetre-scale drawing of posterior part of the skull-base in hypertelorism.

F.M. Foramen magnum.	S.P. Styloid process.
O.C. Occipital condyles.	A.M. Acoustic meatus.
O.B. Os basillare.	F.H. Foramen of Huschke.
C. Crano-pharyngeal canal,	F.S. Foramen spinosum.
inferior opening of.	F.O. Foramen ovale.
J.F. Jugular fossa.	

left, and on neither side is a condyloid foramen present. Each condyle presents two articular facets, this division being even more obvious in the corresponding surfaces of the atlas. The arch of the atlas is incomplete posteriorly.

The foramen magnum is symmetrical and comparatively large, measuring 35 mm. in its antero-posterior and 29 mm. in its greatest transverse diameter. The edge of its posterior segment is unusually thin and sharp. A striking feature of this skull-base is the foreshortening of the squamous portion of the occipital below the highest nuchal line, from the opisthion to the inion measures but 15 mm. The nuchal lines, however, are



## David M. Greig

well developed, and the surface copiously marked by muscular and fascial attachments.

The palate forms practically a plane surface. Its length from anterior nasal spine to posterior nasal spine is 46 mm. and its breadth between the two molar teeth 29 mm., or between the outer alveolar margins 50 mm. (Fig. 26). The palatal processes of the palate bones are well formed, but the pterygoid processes are slight and the pterygoid fossa narrow in its transverse diameter. The palate is neither arched, narrow, nor high, but it must not be overlooked that alveolar processes, and perhaps

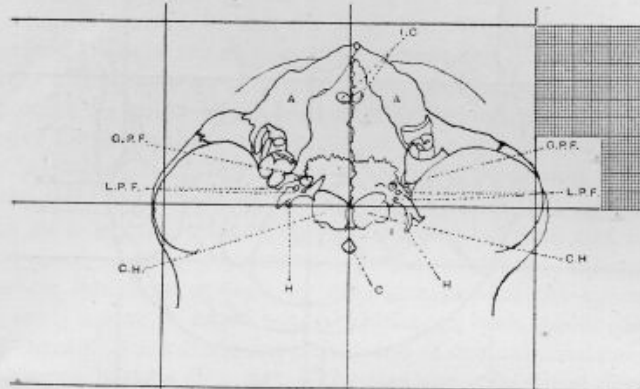


FIG. 26.—Millimetre-scale drawing of the anterior portion of the skull-base in hypertelorism.

- |   |                                   |
|---|-----------------------------------|
| C. Cranio-pharyngeal canal,<br>inferior opening of. | L.P.F. Smaller palatine foramina. |
| H. Hamulus.   | G.P.F. Greater palatine foramen.  |
| Ch. Choane.   | I.C. Inclisive canals.            |
|   | A. Alveolus.                      |

voluminous gums, may have given it a different aspect in early childhood. The premaxillary elements are present, and there is a wide incisive foramen in which the two component foramina are plainly visible.

**The Interior of the Cranium.**—There is no general abnormal characteristic. The internal table is normal in its smoothness and in its quality, and the grooves for the vascular channels are normal in depth and distribution, and the few pin-point foramina opening along the groove for the sagittal sinus have already been noted. There are no digital impressions seen in the radiogram (Fig. 27). In describing the fossæ in detail it will be more convenient to review them from behind forwards.

**Posterior Cranial Fossa** (Fig. 28).—From inspection of the

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exterior of the skull scant accommodation for the cerebellar hemispheres might have been expected, but the reverse is the case, for these fossæ with their steep sides are deep, large, and unusually commodious. The internal occipital crest is broad and the grooves for the transverse sinuses of ample size. The internal occipital protuberance bears no relation to the external occipital protuberance being situated at a higher level by 40 mm.

The lateral half of the superior margin of the petrous portion is sharper, and the recess under it for the sigmoid sinus deeper, than usual. On the left petrous bone the posterior surface shows below the opening of the aquæductus vestibuli a bony deficiency in relation to the termination of the sigmoid sinus (Fig. 29). The dorsum sellæ is broad and strong, and the terminal tubercles of the posterior clinoid processes are large and rounded. On the left side of the middle line the dorsum sellæ is perforated by a foramen, while on the right side a thin plate of bone occupies the corresponding site.

**Middle Cranial Fossa** (Fig. 30).—In comparison with the large recesses for the cerebellar lobes, the lateral recesses in the middle fossa for the temporal lobes of the cerebrum seem small, but they are not shallow as in hydrocephalic skulls. The pituitary fossa is large and forms a cup-shaped depression possessing lateral boundaries. There is no similarity to a saddle as the floor lies below the level of the lateral walls, an arrangement which would prevent an accurate appreciation of the size and shape of the fossa by radiography. In front of the deepest part of the fossa is the internal opening of the cranio-pharyngeal canal. The tuberculum sellæ forms a smooth, scarcely appreciable, fullness in the sulcus chiasmatis which curves upwards to each optic foramen. The transverse concavity of this portion of the sphenoidal body gives the optic foramina the appearance of being situated at an unusually high level (Fig. 31). The anterior clinoid processes are massive and project further back than usual. They closely approach the posterior clinoid processes and have none of the downward inclination found in hydrocephalic skulls. A small middle clinoid process is present on each side. The great wings of the sphenoid are overwhelmed by the size and solidity of the small alæ. Each large wing is narrow, its posterior border runs parallel with its anterior, and its greatest diameter measures but 13 mm. Laterally the right great wing terminates at the lateral extremity of the small wing, but on the left

side it falls 13 mm. short of that point. Each optic foramen is triangular in shape, of large size, and looks upwards and

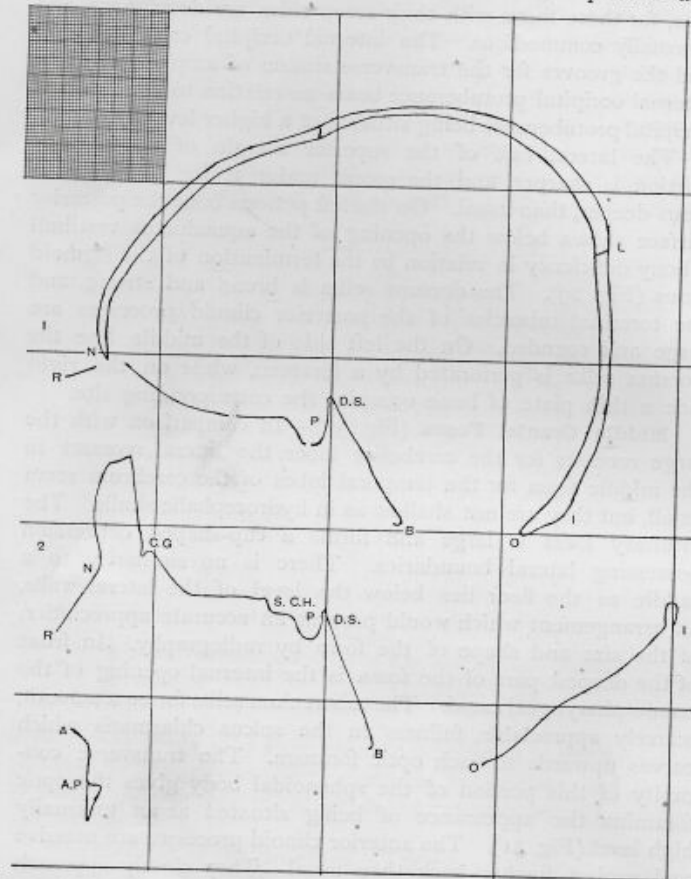


FIG. 32.—(1) Outline sketch of interior of cranium in hypertelorism; (2) Outline sketch of interior of cranial base of a 14-year-old skull for comparison.

R. Rhinion.	O. Ophryon.	S.Ch. Sulcus chiasmatis.
N. Nasion.	B. Basion.	C.G. Crista galli.
Br. Bregma.	D.S. Dorsum sellae.	A. Acanthion.
L. Lambda.	P. Pituitary fossa.	A.P. Alveolar point.
I. Inion.		

mesially rather than backwards, and mesially since the roots of the smaller wings are placed horizontally to each other



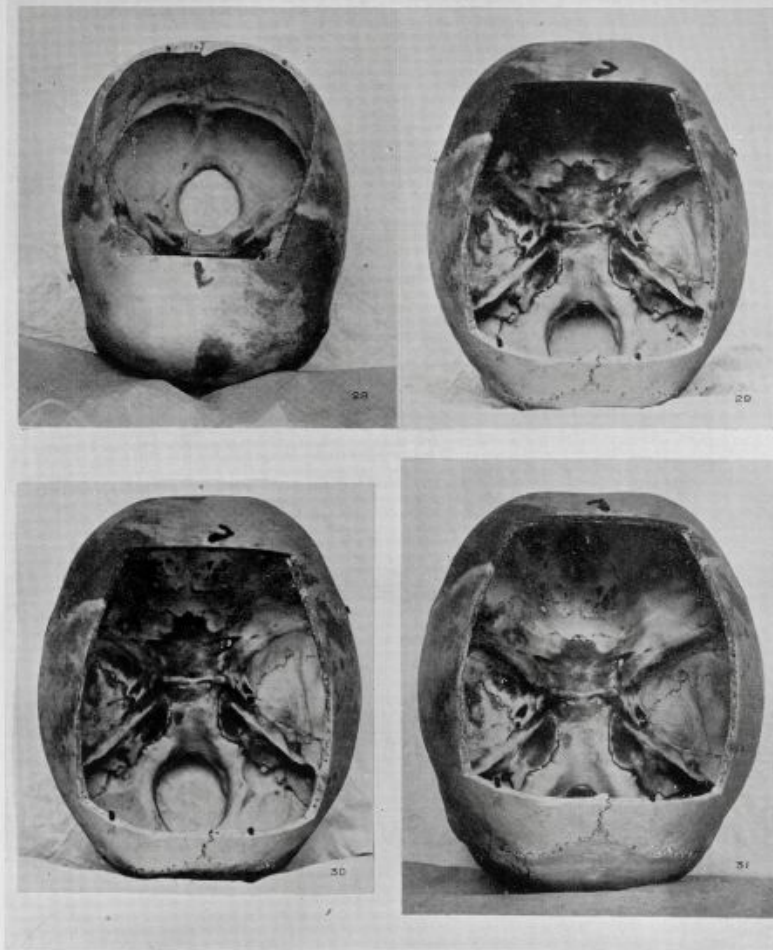


FIG. 28.—Hypertelorism; the skull inverted to show the posterior segment of the posterior cranial fossa.  
 FIG. 29.—Hypertelorism; the anterior segment of the posterior cranial fossa.  
 FIG. 30.—Hypertelorism; the middle cranial fossa. The internal opening of the cranio-pharyngeal canal is seen just over the centre of the dorsum sellae.  
 FIG. 31.—Hypertelorism; posterior part of anterior cranial fossa.

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rather than vertically. The small size of the tuberculum sellæ gives the sphenoidal surface a gradual slope into the pituitary

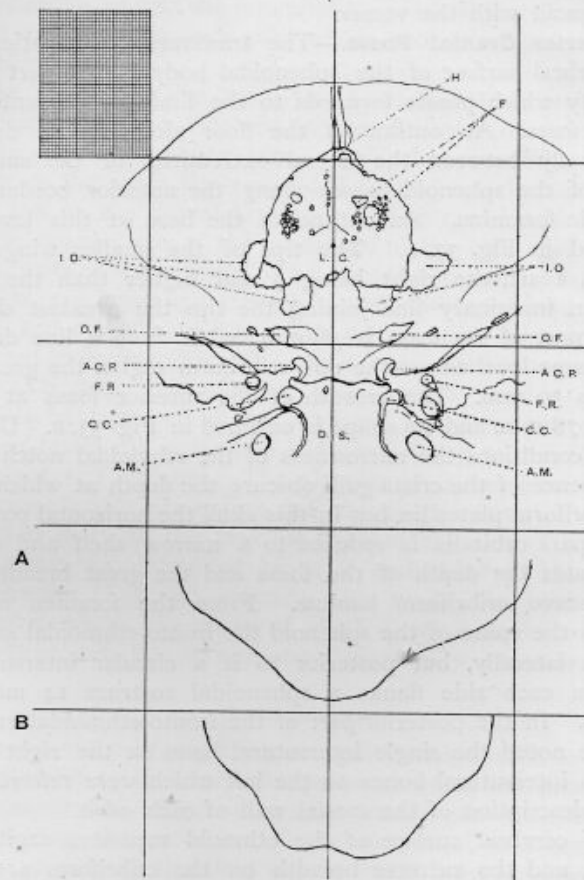


FIG. 33.—Millimetre-scale drawing of the anterior cranial fossa in hypertelorism.

D.S. Dorsum sellæ.	O.F. Optic foramen.
A.M. Acoustic meatus.	I.O. Intersutural bones.
C.C. Carotid canal.	L.C. Lamina cribrosa.
A.C.P. Anterior clinoid process.	F. Foramina.
F.R. Foramen rotundum.	H. Hiatus at nasion.

A. Transverse section across floor of anterior fossa immediately behind posterior edge of great wings of sphenoid.

B. Transverse section of anterior fossa across centre of lamina cribrosa.

fossa, and the absence of a posterior lip to the sulcus chiasmatis lessens the depth of the pituitary fossa anteriorly (Fig. 32).

The tip of the sphenoidal spine is wanting, and its site occupied by a foramen which leads forwards and downwards and finds its exit in front of the junction of the perpendicular plate of the ethmoid with the vomer.

**Anterior Cranial Fossa.**—The transverse concavation of the cerebral surface of the sphenoidal body is but part of a concavity which passes forwards to the limit of the anterior cranial fossa. An outline of the floor along a line drawn transversely between the lateral extremities of the smaller wings of the sphenoid passes along the anterior borders of the optic foramina. An outline of the base at this level is indicated in Fig. 33, A. The tips of the smaller wings lie 96 mm. apart, the right being 3 mm. higher than the left. From an imaginary line joining the tips the greatest depth of this part of the fossa is 36 mm., while from a line drawn at the same level across the mid-cribriform region the greatest depth is 39 mm. The breadth of the anterior fossa at this level is 78 mm. and its shape is outlined in Fig. 33, B. Under normal conditions the narrowness of the ethmoidal notch and the presence of the crista galli obscure the depth at which the flat cribriform plates lie, but in this skull the horizontal portion of the pars orbitalis is reduced to a narrow shelf and even accentuates the depth of the fossa and the great breadth of the concave cribriform laminae. From the foramen which replaces the spine of the sphenoid the fronto-ethmoidal suture extends laterally, but posterior to it a circular intersutural bone on each side flanks a sphenoidal rostrum 14 mm. in breadth. In the posterior part of the fronto-ethmoidal suture is to be noted the single intersutural bone on the right and the two intersutural bones on the left which were referred to in the description of the mesial wall of each orbit.

The cerebral surface of the ethmoid measures sagittally 38 mm. and the extreme breadth on the cribriform area is 40 mm. At the site of the crista galli are five minute foramina lying in line sagittally. They lead to the interior of the thickened anterior end of the perpendicular plate (Fig. 34).

The cerebral surface of the ethmoid may be divided transversely into three portions, a posterior which measures about 11 mm. and is smooth and imperforate, a central which measures about 14 mm. and is the cribriform area, and an anterior which measures about 13 mm. and is smooth and imperforate except for the sagittal row of foramina already mentioned. The



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cribriform portion is the most concave and sagging. The openings in it occur mostly in the lateral part of each lamina. They are most numerous close to each lateral suture where they form fenestræ, and all the foramina are of small size except one placed anteriorly on each side which probably conveyed the naso-ciliary nerve.

**The Mandible** (Fig. 35).—The foetal characteristics of the mandible have been retained in the shortness of the ramus and the obtuseness of the angle. In each ramus (Figs. 36 and 37) above the angle lies embedded an unerupted third molar tooth, and on the lateral aspect of the left ramus a deficiency of bone exposes one of the roots, and of the right ramus two of the roots of these teeth. The mandibular and the mental foramina are large and there is a well-developed mylo-hyoid groove and mylo-hyoid line. The mental spine forms a sharp, backward projection (Fig. 38). As is usual when the mandible projects beyond the maxillæ the anterior portion of the mandible has acquired an upward curve. The carious remains of seven teeth still exist, two fragments being too small to be recognised and replaced. The alveolar processes have undergone considerable absorption but not to the same extent as has taken place in the maxillæ. About the site of the first right molar an unerupted tooth is embedded in the bone. Radiography (Fig. 39) indicates this to be the root of an inverted premolar, the crown of which directed downwards and forwards projects into an almost globular cavity, a corono-dental cyst.

### Conclusions.

Anatomical extravagances are particularly liable to occur along lines of evolutionary change or where in the process of development one region or system passes into another, and remembering the evolution of the face, skull, and brain we can appreciate the threefold factors which may exercise their influence in the development of this part. In other words, we have to consider the soft parts, the bones, and the cranial contents. I think we may exonerate the soft parts. From the absence of defects along the evolutionary lines of union, from the absence of scar tissue, and from the presence of normal muscular activity we may conclude that not herein lies the provocative agent.

In the skull the most striking peculiarity is the size of the anterior nasal aperture with the great breadth of the cribriform

plates, as during life the most striking peculiarity was the hypertelorism. Could it be that an increase in breadth of the fronto-nasal region has been determined by an overgrowth of the horizontal plate of the ethmoid pushing aside the two halves of the frontal bone? To admit this would surely be to ascribe to these thin and delicate plates a power beyond their capabilities, especially as by the under-development of the rest of the ethmoid, the cribriform laminae are deprived of a material support. So here we may leave this matter of the cribriform plates for a moment and turn our attention to the next most striking feature, which is the small size of the great wings of the sphenoid and the disproportionate largeness of the small wings. This was at one time accepted as an embryonal characteristic; but, as Professor Fawcett<sup>10</sup> points out, that is accurate only in relation to that part of the sphenoid which is developed in cartilage, and he puts the order of ossification of this region as follows: (1) internal pterygoid plate; (2) ala temporalis (great wing); (3) orbito-sphenoid; (4) processus alaris; (5) sphenoidal turbinate bones. Certainly in the chondro-cranium the smaller wings are much larger than the greater wings (Fig. 40). Ossification, however, occurs earlier in the ala temporalis (cartilaginous portion of the great wing) than in the orbito-sphenoid, but the accession to the greater wing of the part developed in membrane soon reverses the relative sizes of these two portions of the sphenoid.

This observation by Professor Fawcett is for our present purpose one of very great interest. In the general review of the middle fossa of the cranium it was pointed out that the great wings of the sphenoid were overshadowed by the small wings, and that the great wings did not extend laterally on the right side beyond, and on the left side even as far as, the lateral terminations of the small wings. It was noted that the anterior and posterior borders of the alisphenoids were almost parallel and that their greatest breadth was but 13 mm. Laterally, it is true, each alisphenoid is supplemented by an epipteric bone, but that brings in another question which I shall deal with presently. Obviously the small size of the alisphenoids cannot explain the deformity, for it is not difficult to select a skull, especially among those of mentally deficient persons, in which the alisphenoids are no larger than they are in this case, yet these skulls do not present the peculiarities of hypertelorism.

Ossification of cartilage begins in the pterygoid about the

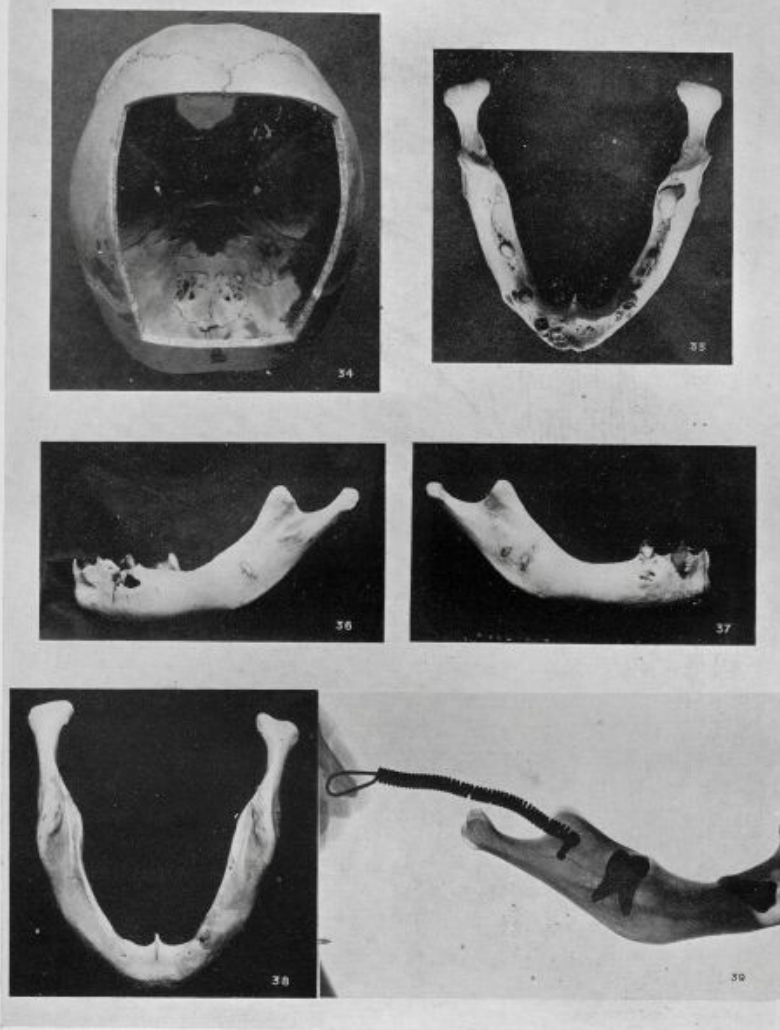


FIG. 34.—Hypertelorism; skull inverted to show the anterior segment of the anterior cranial fossa, especially the size and shape of the laminae cribrosae.  
 FIG. 35.—Hypertelorism; the mandible viewed from above.  
 FIGS. 36 and 37.—Hypertelorism; the left and right halves of the mandible viewed laterally.  
 FIG. 38.—Hypertelorism; the mandible viewed from below.  
 FIG. 39.—Hypertelorism; radiogram of right half of mandible to show an unerupted molar tooth and the inverted posterior premolar embedded in the bone.



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fifty-third and in the ala temporalis about the fifty-ninth day, that is in the eighth week. It must not be assumed of course that a bone defect necessarily dates from its ossification; the defect may be, in all probability is, in the scaffolding in which the bones form, that is in the chondro-cranium. But it can be assumed that as far as defect in architecture is concerned that defect becomes stereotyped once the parts are set in bone. The question then arises, Does the portion of the sphenoid which is developed in cartilage show signs of abnormal development?

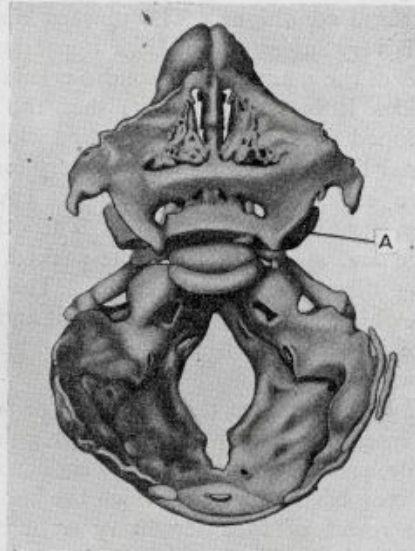


FIG. 40.—Chondro-cranium 40 mm. human embryo (63 days).  
(After Macklin.)

A. Ala temporalis (great wing) of the sphenoid.

The answer to this is certainly in the affirmative. The very presence of the cranio-pharyngeal canal suggests such a possibility. This canal lies between the basisphenoid and the presphenoid. It contains the naso-pharyngeal stalk of the pituitary which is formed by the seventh week and becomes elongated as the body of the sphenoid insinuates itself between the pharynx and the forebrain, and normally by the ninth week the stalk has disappeared and the canal closed. Further, its external opening lies 5.5 mm. posterior to the nasal septum and not under cover of the wings of the vomer as is usually

reported when the canal is present.<sup>11</sup> \* Reconsidering the base of the skull in this light the nasal septum seems to be too far forward, for it leaves no less than 13 mm. of sphenoidal body exposed between the nasal septum and the sphenoccipital junction. Abnormal ossification is further shown by a little irregular mass of bone which lies at the root of each mesial pterygoid plate. This is the chondral portion of the pterygoid, the earliest portion to undergo ossification. Then turning to the cranial surface of this region the great size of the pituitary fossa has already been noted and the massive dorsum sellae is reminiscent of foetal conditions. The upward direction of the optic foramina is very suggestive of their appearance as figured in some pictures of the mammalian chondrocranium.

On both sides it is seen (Fig. 31) that the parts of the sphenoid developed in cartilage are packed closely together. The carotid canal lies so close to the pituitary fossa that only a plate of bone separates the two and the cavernous sinus must have been very short and narrow. The other foramina, rotundum, ovale and spinosum are close-packed, the foramen spinosum being disproportionately large. The lingula, that curious process which guards the carotid canal and is developed from a centre of its own, is well marked and all but touches the tip of the petrous bone.

Here then I think we have the *fons et origo mali* of hypertelorism. An interference with development, going back early in embryonic life, present certainly in the chondrocranium and perhaps in the mesoblastic tissue in which the chondrocranium is laid down, a result of that hereditary want of balance in development, which in families marked by Nature for extinction, brings forth the physically and mentally maimed and defective individuals whom we stigmatise as "degenerates."

This is my hypothesis of the cause of the defect, but before passing to consider if it can explain the other features peculiar to this skull let me say a word about intersutural bones. I believe that intersutural bones are apt to occur wherever there is the combination of open sutures and strain. In some positions intersutural bones are so common as to be almost normal, and under normal or abnormal conditions they develop bilaterally and symmetrically because the conditions of growth and of brain pressure are bilaterally equal. Disturb that equanimity and a unilateral production of intersutural bones is the result.

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To return then to the consideration of this skull. The disturbance of growth is entirely in the prechordal part of the cranium, and therefore particularly related to the nose and eyes. The initial lesion is a limited one, but its effects are far reaching. It must not be forgotten that while chondrification and ossification are proceeding in the base of the primitive cranium, and to a much greater extent during ossification of the membrane-developed superstructure, skull-moulding is modified by the growing brain, and it is the telencephalon and the thalamencephalon which will be specially involved. That part of the brain which develops behind the dorsum sellæ, the mid-brain, the hind-brain, cerebellum, and pons, is not affected, at any rate not to any appreciable extent. The immediate result of the interference with the development of the ala temporalis is that the brain has not in that part sufficient room for expansion and it forces itself laterally. Interference with growth in the region of the pterion results, as is seen in oxycephaly and many other congenital deformities, in a retention of the great depth of the occipital chondrocranium which is found in the embryo,<sup>13</sup> and this entails a vertical position of the supra-inial portions of the occipital bone and generally results in brachycephaly. In such circumstances no strain is thrown on the lambdoid suture, therefore no intersutural bones are developed. The want of backward development of the occiput, though not inconveniencing the cerebellum or posterior lobes, gives the growing brain a forward impetus. As a result of shortcomings in the formation of the bones and increased lateral cerebral pressure numerous and large intersutural bones have formed at the asterion and pterion, and perhaps flattening or opening out of the fissures of Sylvius may have determined the unusual thickness of the posterior borders of the smaller sphenoidal wings.

As far as the brain is concerned the interference with its backward and lateral expansion has thrown the stress chiefly on the prosencephalon. Posterior to this, the growth of the great ganglia of the brain-base has proceeded uninterruptedly and retention of their functions has been assured. Therefore the individual develops normal ambulatory powers, normal sensation, and is of sufficient physique; though one should not be surprised to find a disturbance of the tectal autonomic centres which might find expression in want of full somatic growth and in the presence of acrocyanosis. But anterior to the dorsum sellæ, deformation of the brain and the pressure on



it have interfered with the development of the higher cerebral functions. In the growth of the pallium and neopallium the nerve cells or the grouping of them have been interfered with, or the complicated system of connecting tracts between the higher ganglia or even between the two cerebral hemispheres may have been interrupted or imperfectly evolved. The result would be imbecility, using that term as intermediate between idiocy and a higher grade of mental deficiency. The initial defect in the skull which I have described has fully developed in the embryo by the third month but the great pallial growth only commences then. The volume and bulk of the brain has been but little interfered with though the functions have been materially affected.

At an early period of embryonic life, even at the sixth week, the anterior cerebral vesicles bulge from the front part of the prosencephalon, while the fronto-nasal process forms a broad belt of tissue at a lower level in the middle line. At this time too the chondro-cranium forms, and as soon as the abnormal smallness of the ala temporalis makes itself felt pressure will be produced anteriorly. The subsequent want of room for lateral expansion and the continuing pressure from behind would account for the retention of the foetal facies. Some moulding of the chondro-cranium would take place, and the jugum sphenoidale be rendered or remain concave from side to side. For the same reason the tuberculum sellæ would be poorly developed, and the sulcus chiasmatis would not assume a backward direction but remain directed upwards, as also would the internal openings of the optic foramina. The pressure which renders or keeps concave this portion of the sphenoid must act even more decidedly further forward, and the horizontal plate of the ethmoid, that is the cribriform laminae, would form a concavity and be extended in all directions but particularly in breadth. Undoubtedly a strain occurs at the junction of these bones and this strain would be bilateral and symmetrical, and so in the spheno-ethmoidal suture, in the spheno-frontal suture, and in the fronto-ethmoidal suture, there is a symmetrical development of intersutural bones. But another peculiar result has been produced, namely, that in the spreading out of the cartilaginous nasal capsule, or by a centrifugal increase in size during ossification, the formative material of the ethmoid is used up in the abnormal size of the cribriform laminae, and there have come into being neither

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cristi galli nor ethmoidal labyrinths. Finally the concavation and broadening of the spheno-ethmoidal cerebral surface could not have occurred without traction, and this would tilt upwards the anterior portion of the nasal capsule and bring about the vertical axis of the nasal bones sagittally; while the orbits retaining their foetal lateral deviation would, by traction in the horizontal plane, ensure nasal bones of an unusual breadth and a strain which would produce in the internasal suture an intersutural bone.

The importance of the sphenoidal bone and its relation to cerebral development is nothing new. Three-quarters of a century ago, albeit more than ten years after the *debacle*, my illustrious predecessor, our first Conservator, Dr Knox,<sup>12</sup> noted its value and briefly referred to the skull of an adult male congenital imbecile in which a sphenoidal deficiency was the sole osseous peculiarity. Under Sir John Struthers' ægis I feel I am following worthy footsteps.

It may be asked: Why add a new name, another variety, to the already copious varieties of cranial deformities? Can not hypertelorism be classed under one of the recognised varieties? The answer is: It can not. Hypertelorism is an entity having a definite origin, a definite course. It is dependent on a growth-disturbance of a minute portion of the embryonic cranial tissue, from which consecutively certain deformities are produced. The deformities may vary in degree according to the degree of primary defect, but they are otherwise definite and fixed, and nothing else can simulate them, and the combination can only be caused by this one lesion for which the fortunate acquisition of this skull has enabled me to submit an explanation.

### Addendum.

As the title of this lecture infers, I am unaware of any description of hypertelorism as an entity or even as a leading feature, and I do not know of any published description of a similar skull. Perhaps the following may not be irrelevant.

In 1890, as a variety of plagiocephaly, Fridolin<sup>14</sup> described the skull of a boy aged 3 months. It was asymmetrical, high and short, and the right side flatter than the left. The frontal and parietal eminences were absent from the right side, and the upper two-thirds of the right half of the coronal suture were synostosed. In my opinion the partial synostosis and the absence of the right parietal and frontal eminences are related

to each other, the synostosis having prevented their development, and, an unwonted strain being thrown on the anterior fontanelle, a large intersutural bone has been the result. This part of the deformity is not uncommon, and has no particular relation to hypertelorism, but the other part of the deformity present has; and the presence of two defects in the skull need excite surprise no more than the coexistence or multiplicity of congenital defects elsewhere.

The right orbital cavity is higher and narrower than the left, and the distance between the eyes is in consequence increased. The right orbital index is 155, the left is 84. In the right temporal region the great wing of the sphenoid is much narrower than in the left, the measurements being respectively 3 mm. and 11 mm. The right zygomatic bone is higher than the left, and the zygomatic arch is shorter. The right side of the root of the nose is 54 mm. from the acoustic meatus, the left is 69 mm. from the corresponding point on the left side. The right zygomatico-maxillary suture is partly synostosed; the infra-orbital suture is still open. The right nasal bone is absent; the left is very broad and extends to the right over the middle line; \* it measures at its root 9 mm. The nasal process of the right intermaxillary bone to which the inferior concha articulates by its anterior extremity is separated from the maxilla by a fissure 3 mm. deep—the suture intermaxillaris facialis. A fissure still separates the two intermaxillary bones. The interpalatal suture inclines to the left, and the long axis of the foramen magnum to the right. The lower root of the smaller wing of the sphenoid is absent from the right side, so that the optic foramen is not closed up, and on the same side the anterior and middle clinoid processes are synostosed. There is no left middle clinoid process which of course is normal for an infant of this age. The right side of the mandible is less developed than the left, a derangement, partly developmental and partly due to imperfect musculature which would accompany the right-sided osseous defect.

It is obvious that synostosis of part of the right side of the coronal suture could not explain all these defects which are entirely unilateral. It is not difficult from what we have seen in hypertelorism to visualise the right side of the skull Fridolin so minutely describes. Further, if one can imagine the deformity as bilateral, the similarity to hypertelorism will be at once

\* Is it not that the two nasal bones are fused?



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recognised and the fault ascribed to the sphenoid, especially that portion of its greater wings which is developed in cartilage. The explanation I have given of hypertelorism makes a unilateral affection not only possible but probable.

In the osteoclastic activity which has resulted in so great alveolar absorption there is probably nothing to wonder at. A septic periodontitis, in a mentally-deficient person, incapable of being taught dental hygiene and nourished for the most part on soft, digestible, and easily-swallowed food, is certain to tend towards an osteoporosis rather than to provoke an osteoblastic reaction. There is other evidence, for example, in the osseous walls of the acoustic meatus and at the apex of each petrous bone, of unstable osseous tissue and perhaps in the imperfectly-developed brain cerebral control of osseous growth may have been abnormal.

Maggi<sup>15</sup> and Waldeyer<sup>16</sup> found the cranio-pharyngeal canal present in 40 per cent. of anthropoid apes, and it has been demonstrated by other investigators as present in rabbits, cats, and other animals with considerable frequency. It would be surprising if it were not occasionally found in the human subject, and in most text-books of human anatomy the occasional existence of a patent cranio-pharyngeal canal is referred to.

It is common to find in a full-time foetus or in the newly-born child the site of either end of the cranio-pharyngeal canal indicated by a blind canal or by a funnel-shaped impression in the bone. Statistics of frequency vary much in accordance with the age of the skulls examined, and it is significant that the occurrence of a patent cranio-pharyngeal canal diminishes with age. Some facts as to its persistence have been usefully collated by Haberfeld.<sup>17</sup> Landzert,<sup>18</sup> who was the originator of the name "cranio-pharyngeal canal," says he found a complete canal present in 10 per cent., and Le Double<sup>19</sup> in 9 per cent. of newly-born children. If the numbers of the skulls examined by the authors quoted by Haberfeld be added together, excluding Le Double's observation of 100 skulls under 3 months of age, a complete cranio-pharyngeal canal is found twenty-nine times in 6281 skulls, or in rather less than 0.5 per cent. of presumably otherwise normal crania of all ages. As regards sex incidence opinions differ. Le Double found the canal present eighteen times, of which 11 were males, while Casselli, in a much more extended investigation, found it present twelve times, of which only 3 were males. The remains of the cranio-pharyngeal

canal must not be confused with the one or two minute canals, venous channels to the interior of the bone, which are not uncommonly found in the sagittal plane but a little distance in front of the cranio-pharyngeal canal.<sup>20</sup>

Breschet<sup>28</sup> observed a canal through the body of the sphenoid in a much deformed female foetus, the subject of ectopia cordis and encephalocele. Fridolin has described one foetal skull<sup>21</sup> in which a patent cranio-pharyngeal canal was in association with left coronal and right lambdoid synostosis, and a right hare-lip, and another<sup>22</sup> in which a more extensive cranial osseous defect existed. The condition of the soft parts was not known, but of course the existence of the canal would mean persistence of hypophyseal stalk though that might have been reduced even to a fibrous cord. Bitot<sup>23</sup> describes it as patent in a case of complicated superior median hare-lip. The combination of patent cranio-pharyngeal canal with cleft palate and hare-lip calls to mind Lichtenberg's case,<sup>24</sup> where a female child born with that deformity had a bluish-red tumour, "the size of a small fist," dependent from the mouth. It was an autochthonous tumour of hypophyseal tissue. A ligature was passed round the pedicle which attached the tumour to the back of the pharynx, and on cutting through the pedicle reddish cerebro-spinal fluid escaped from the distal portion. The child died of convulsions three days later, and the pedicle was then found to pass through the sphenoid bone to the pituitary fossa where it was attached to an oval tumour of grey matter covered by pia mater. The lateral ventricles were dilated and there was a congenital defect in the heart. A happier result followed Lücke's operation in the case described by Sonnenburg<sup>26</sup> of a 5-days-old child. The epignathus depended from the skull base through the cleft hard and soft palate and protruded from the mouth. It was of a teratomatous nature, and Sonnenburg deprecated its being a spheno-pharyngeal hernia, and doubted if it were connected with the pituitary through a cranio-pharyngeal canal. Windle<sup>25</sup> has described as a variety of epignathus a somewhat similar though larger tumour associated with absence of the septum nasi. The tumour attached by a fibrous stalk which passed through the cranio-pharyngeal canal overflowed through the mouth and nasal apertures. Such examination as its decomposed state admitted proved it to be heterochthonous, and it may have been a teratoma. The further pursuit of these

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congenital defects leads to the varieties of anterior craniochisis which are often but exaggerated examples of patent cranio-pharyngeal canal incompatible with life, and though developmentally are not clinically interesting.

In normal skulls there is considerable variation in the cribriform laminae, their length, their breadth, the depth at which they lie, and the proportion the crista galli bears to them. It is many years since Sir John Bland-Sutton<sup>26</sup> noted the absence of the crista galli in the chimpanzee, and Duckworth<sup>27</sup> states that the crista galli does not exist in the Simiidae as a rule, but complete absence in man is extremely rare. It is generally associated with absence of the cribriform laminae. Thus Fridolin<sup>28</sup> has described a skull in which absence of the cribriform laminae and of the crista galli was associated with absence of the premaxillae, the vomer, and the perpendicular plate of the ethmoid. The corresponding cerebral portion is not described, probably because it was not available. Nevertheless, variations in the cribriform laminae are generally associated with defects in the rhinencephalon. Schmidt<sup>29</sup> mentions a 29 cm. foetus which he had examined, in which he found absence of the crista galli and the horizontal plate of the ethmoid. The dura mater simply passed intact over this area, but there was no sign of an anterior perforated spot in the brain nor of olfactory bulbs or tracts. Morell Mackenzie<sup>30</sup> quotes from Pressat<sup>31</sup> the report of a post-mortem examination on an adult who had congenital anosmia. No trace of olfactory bulbs or roots could be found but the crista galli was present, as was also the horizontal plate of the ethmoid but it was imperforate. Probably a similar condition was present in Levi's case,<sup>32</sup> but though the bulbs and tracts were absent from the brain of a female idiot aged 3 years the skull is not described in detail. The crista galli was not absent, though the right cribriform plate was in the case described by Selenkoff.<sup>33</sup> The depth at which the cribriform plate lies depends on the size and presence of the frontal air sinuses which, in my case of hypertelorism, had not developed at all. It is obvious that this osseous region develops in relation to the olfactory bulbs, and that the bulbs modify the bones except in such rare circumstances as Schmidt's case,<sup>29</sup> where the intracranial connection of a frontal tumour had modified the anterior part of one cribriform lamina and cramped the development of the corresponding olfactory bulb. I have failed to find any reference



to so large a horizontal ethmoidal plate as I have described in hypertelorism, and it is plain that this extraordinary development is in no way due to any anatomical peculiarities of the olfactory bulbs.

There is little doubt that in its fully-developed state hypertelorism is a rare deformity, but minor manifestations of it are frequently to be observed. Of hypertelorism, as of so many abnormal or diseased conditions, a better comprehension may be obtained from the careful study of one fully-developed example than from the consideration of many of less degree. Its recognition as a particular defect will withdraw from among other cranial deformities cases which have hitherto but confused their appreciation, and its isolation will facilitate the classification of those which remain. These and other reasons referred to in my paper seem to me to justify the admission of hypertelorism as a definite variety of cranio-facial deformity.

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