On oxycephaly / by H. Morley Fletcher.

Reprinted from

The Quarterly Journal of Medicine

April 1911 (*Vol. 4 No. 15*)

ON OXYCEPHALY

By H. MORLEY FLETCHER

With Plates 30–38

The cranial deformity now generally known as oxycephaly is also described under various appellations, such as acrocephaly, 'Thurmschädel,' 'Spitzkopf,' tower or steeple-head, 'turritum caput,' 'tête à la Thersite' (Hamy). The condition does not appear to have attracted attention, except as a curious malformation, until the visual defects so frequently associated with it led to investigations on the part of ophthalmologists. Thus we find that all the earlier accounts of this condition, and there are many of them, appear in the various ophthalmological journals of Europe and this country.

The almost invariable impairment of vision with its frequent accompaniment, optic atrophy, was first definitely recognized and described by v. Graefe in 1866, Michel in 1873, and v. Hirschberg in 1883, and since then a large number of cases of this type, as well as of other cranial deformities associated with optic atrophy, have been recorded, notably by v. Hirschberg, Enslin, and Patry. The bibliography shows that many cases have been recorded; the number cannot be accurately given as some are doubtful and others are re-quoted, but it is probably between eighty or ninety. The condition is not very rare and it is very striking, yet it is surprising that in this country it appears to have met with little recognition by clinicians until quite recently.

My own attention was directed to the condition by a case I met with about eight years ago, at the East London Hospital for Children, in conjunction with my colleague the late Mr. Hancock, and since that time I have been endeavouring to investigate the subject further, as it presents points of very considerable interest.

Clinical Aspects.

The cardinal signs are the characteristically shaped skull, exophthalmos and impairment of vision, sometimes amounting to total blindness, associated with changes in the fundi oculorum.

The aspect of the patient is very striking, the great height of the forehead sloping gradually upwards to the vertex with feebly marked superciliary ridges. The vertex of the skull appears pointed instead of flattened or rounded, and a ridge, or bony prominence, is sometimes felt in the region of the bregma. The hairy scalp may be raised above the normal level and present the appearance of

being perched on the top of a cone. Viewed laterally, the ears appear placed on a lower level than normal. The temporal ridges and depressions are feebly marked.

The eyes. Exophthalmos is present in all the well-marked cases. It is sometimes extreme, as in Cases I and VI, and dislocation of the eyeballs in front of the eyelids occasionally occurs. Failure of closure of the eyes, especially during sleep, may lead to lachrymation and conjunctivitis as in Graves's disease. Not infrequently the protrusion of the eyeballs is unequal on the two sides. Divergent *squint* is common and gives a peculiar appearance to the bulging eyes: occasionally the squint is convergent. *Nystagmus* is present in the majority of cases.

Other facial characteristics. The nose may be well formed, but in several cases there was a definite deflexion to one side. That shown in Plate 30, Fig. 1 (Case I), is probably largely the result of an accident in quite early life. These photographs show well a marked flattening or depression in the malar and supra-maxillary region, which is very characteristic, though not so commonly present in the juvenile as in adult patients.

The complexion tends to be muddy, sallow, and dark, especially in the older patients. The hair was dark in all the cases I have seen.

They are usually mouth-breathers; the open, gaping mouth adds to the grotesque appearance and gives it a stupid vacant expression. In many cases the palate is shortened and extremely highly arched, as in Cases I and III. The teeth are liable to early caries, the incisors are prominent. The last upper molars may remain unerupted, as in Case I, owing to the shortening of the superior maxilla.

Intelligence. This is unimpaired in quite a large proportion of cases, and may be above the average. It is probable that in cases with a mental development below the average this may be due to the ocular defect which develops in early life and produces a backwardness from 'deprivation', to use Ireland's term. One of my cases (Case III) showed a remarkable degree of intelligence: finding that he was unable to distinguish letters he speedily taught himself to read the blind alphabet. Case I never went to school on account of his visual defect, but taught himself to read and write at home with his father's help. Potts states that in examining thousands of mentally defective patients he met with only two cases of oxycephaly.

Special Senses.

Vision. This is occasionally unimpaired, but in the vast majority of cases sight is very defective. This depends to a considerable extent on the age of the patient: the younger the case the better the vision. In the adult cases the ability to read type is rare; perception of large objects is the usual condition. Complete blindness may result, but it is difficult to give accurate figures as to the proportion of cases in which this occurs.

Intra-ocular changes. The most constant abnormality found is *optic atrophy.* The disks appear greyish, bluish, or ivory white with somewhat irregular edges. The arteries are thin, the veins dilated and tortuous. Optic atrophy was present in six of the seven cases to be described later. In one of Dr. Hutchison's cases included in this series the disks were reported as pale, in one atrophied, and in the third the atrophy was of secondary type. In some cases 'choked disk' has been met with in the early stages of the condition of oxycephaly. In forty-two cases collected by Enslin the atrophy was post-neuritic in thirty-six; in only two of these was it regarded as primary. Friedenwald describes post-neuritic atrophy in nine out of twelve cases. Too much reliance must not be placed on these figures as an indication of the frequency of optic atrophy in oxycephaly, seeing that most of the recorded cases have sought advice on account of failing eyesight. They suffice to show that it is a very frequent occurrence, although not an invariable one. Occasionally pigmentary changes in the retina in the neighbourhood of the disk are met with. Errors of refraction, particularly myopia, are common.

Smell is often completely lost. (Meltzer found this in twelve out of twenty cases quoted by Merle.) It was impaired or lost in four out of the seven cases. *Hearing* usually unaffected. *Taste* very rarely affected. The data with regard to these points are not satisfactory, as in many cases they have not been recorded.

Other Malformations or Abnormalities.

The following have been described in a few cases: Malformations of ears, elbow and shoulder joints, fingers; webbing of toes. Tucker recorded in 1904 a case of oxycephaly in an Indian, in which he noticed a peculiarity of the elbow-joints, which could not be fully extended. I have found this abnormality present in one (Case I) of my cases, and in one of Dr. Hutchison's (Case V). The elbow-joint resists full extension (Plate 31, Figs. 5 and 6). When extended as far as possible, the forearm is deflected outwards. The inner condyle appears to be unduly prominent. This peculiarity requires much fuller observation before any opinion can be given as to its frequency or significance. More will be said about it when discussing the pathology of the disease. In one of these cases (Case I) there is limitation of movement in an upward direction in both shoulder-joints.

Dr. Hutchison and I have met with a very curious malformation of the fingers in two cases (Cases I and V). The first phalanx of each thumb is somewhat thickened, flattened, and sharply curved outwards, with its concavity to the radial side. The little finger in both these cases is curved inwards.

Course.

An analysis of the recorded cases leads me to the conclusion that they may be divided into three groups. In the *first*, exophthalmos and the deformity of the head are definitely present at birth, and in some of these cases total

blindness. These may be classed as congenital cases; they are comparatively few in number, and form but a small proportion of the total. In a *second* group may be placed the cases in which during the first few months of life changes in the shape of the head develop, and the eyes are noticed by the parents to be 'large'. In the course of the first two years these signs become more marked and are attended by gradually increasing impairment of vision. The exophthalmos and the lofty shape of the head become increasingly evident as growth proceeds. The *third* group includes those cases which appear to be quite normal for the first few years of life and the earliest signs may not appear before the second to the sixth year. In these cases an increasing visual defect is usually the first symptom. Vision may remain unimpaired till the fifth to sixth year.

It is interesting to note that in some cases the onset of the condition appeared to date from a fall, or a blow on the head. Patry records two such cases in which this occurred at the age of four years. (Compare Dr. Hutchison's case, VII.)

A consideration of the recorded cases leads me to agree with Patry that the ocular defect begins in the first five years. The impairment of vision is progressive and sometimes ends in total blindness, but in most cases perception of light or of large objects remains. Fair vision may remain with extreme exophthalmos and very definite optic atrophy as in Case I.

As growth proceeds, the lofty shape of the head becomes increasingly evident and exophthalmos develops. Headache is a very common symptom, and may be very severe. In one of my cases it was frontal and vertical. It is often occipital. Vertigo is less frequently met with. Fits have been recorded in a few cases in young children, but it is difficult to be certain as to how far these fits are connected with the cranial condition.

Duration. As far as the evidence serves there is nothing to show that oxycephaly shortens life, but it is a noteworthy fact that few, if any, typical cases have been recorded in patients over fifty years of age.

Predisposing Causes.

Race does not appear to have any influence. In this series of seven cases, four were Hebrews; this high proportion is probably explained by the fact that the cases were drawn from hospitals in the East End of London attended by large numbers of this race. The recorded cases include most of the European races.

Syphilis and *Rickets* cannot, in the writer's opinion, be regarded as important factors, though some writers (Meltzer and Potts) consider that rickets plays an essential part in the pathology of oxycephaly.

Sex. This may be said to be the only important predisposing factor, as the condition is much more common in males than females. Patry gives a series of sixty-four cases in which only seven were females (this series includes some cases which were not true cases of oxycephaly).

Heredity is very rarely a factor. v. Hirschberg records a case whose maternal

grandfather was similarly affected. Weiss and Brugger mention two oxycephalic brothers seen by Oeller. Hanotte states that Hamy met two oxycephalic sisters. The mother of one of my cases (Case I) had a malformed head, exophthalmos, and defective vision.

Morbid Anatomy.

Almost the whole of our knowledge of the morbid anatomy of oxycephaly is derived from the study of dried skulls in museums, the histories of which are almost without exception unobtainable. Very full investigation of the special features of the oxycephalic skull has been made, notably by Hanotte, Patry, Enslin, and others, and the peculiarities found in this condition are constant and characteristic. Through the kindness of Professor Keith I have been able to examine the specimens in the museum of the Royal College of Surgeons, which contains several good examples. Very few, probably not more than three, cases of oxycephaly have been recorded with a post-mortem examination.

In this country Power (1894) recorded a case described as oxycephaly in an infant which lived four weeks, the head of which is preserved in the museum at St. Bartholomew's Hospital, and in 1901 Carpenter described another case, also in an infant, the head of which is in the museum of the Royal College of Surgeons. It is very doubtful, in the writer's opinion, whether either of these cases, particularly that of Carpenter, should be classed as oxycephaly, but rather as foetal monstrosities, as there was no evidence of premature synostosis.

The dried skulls which have been examined belonged mostly to adults, and present the following features:—There is a great increase in the vertical height due to the alterations of shape of the vault. The superciliary ridges and frontal prominences are absent or much reduced and the frontal region rises steeply upwards to the bregma. The frontal and mastoid sinuses are often absent. The temporal fossae are shallow. The characteristic abnormality found is the evidence of premature synostosis of certain sutures. Of these the coronal and sagittal sutures are chiefly involved. The coronal suture is generally completely closed, but sometimes is found to be ununited for a short distance, 2–5 cm., on each side of the bregma. The sagittal suture is usually synostosed. The bregma is often marked by a distinct prominence or bulging. The metopic suture is synostosed. Other sutures are frequently involved, but to a much more varying degree than those already mentioned, but generally speaking those of the vault are the ones most affected. The orbits present striking features. The depth is much reduced, the external part formed by the greater wing of the sphenoid appears to have been pushed forwards. The orbital axes are very oblique downwards and outwards.

The brain. Papillant investigated the condition of the brain in a case of oxycephaly. Bourneville's case was not a case of pure oxycephaly. In Papillant's case the convolutions, more particularly of the lower portion of the brain, showed obvious signs of pressure, while the reverse was manifest in the

upper part of the frontal and parietal lobes. The convolutions with an antero-posterior direction appeared to show arrested growth and were very difficult to separate. The horizontal branch of the Sylvian fissure was very short.

The superior maxilla is very poorly developed, as well as the lateral walls of the nasal chamber, especially in the vertical direction. The nasal septum is generally deviated, probably in part as the result of these changes and also owing to a pushing down of the base of the skull (basi-sphenoid and mes-ethmoid). The total length of the superior maxilla is reduced in proportion to the anterior posterior diameter of the skull.

Some writers have described a narrowing of the optic foramen, but the evidence appears to be against this. The hard palate is sharply arched. The antra may be rudimentary. At the base the condyles are sometimes less prominent than normal. The basilar process is much shortened. The cranial fossae show great changes in size and shape; they are deepened and widened. The middle fossae exhibit this to a striking degree, and this is well shown in the appended skiagrams. The sella turcica is widened and deepened in its central portion. The inferior maxilla does not present changes that can be regarded as characteristic. On holding an oxycephalic skull to a bright light the bones of the vault and of the fossae appear more transparent than those of a normal adult skull, and the so-called 'digital markings' to be referred to later can sometimes be faintly distinguished by transillumination in this way. Measurements of oxycephalic skulls have been very fully made in long series of cases by several writers, particularly Patry, and reference should be made to his work for information on this point.

X-ray appearances. Another method of examining the changes in the skull is by means of the X-rays. Grunmach (v. Hirschberg and Grunmach), Bertolotti, Dorfmann, and others have described the appearances met with which generally confirm in the living subject the existence of the changes already described in dried skulls. All those who have investigated cases of oxycephaly by this method have noticed certain abnormal features which may be regarded as constant. The structure of the bones of the skull, especially those forming the vault, instead of presenting a fairly uniform density in the skiagram show certain markings or depressions resembling a coarse network, and giving the shadow of the bone a dimpled appearance.

These areas, or 'digital markings' as they are called, are most obvious in the frontal region, although they may be clearly visible over the whole vault and in the basal fossae, where they differ from the appearances in cranio-tabes in that they are more diffuse, larger, more numerous, and less sharply defined. These 'digital markings' are occasionally seen in apparently normal skulls, but never, so far as the writer can ascertain, in the same degree as in oxycephaly. They have been generally attributed to pressure exerted by the brain on the skull, and the markings are regarded as corresponding to the convolutions. This explanation does not appear to the writer to be entirely satisfactory, as, in the first place, in another condition, hydrocephaly, the pressure

exerted on the cranial bones does not usually give rise to a similar appearance. For purposes of comparison a skiagram was taken from a patient, aged 52, suffering from effects of old hydrocephaly, and examined. In this the bone presented a much more uniform structure, and in spite of the reduction of thickness the digital markings were absent; the examination made of skiagrams of juvenile hydrocephaly showed exactly the same appearances. Secondly, it is difficult to accept without further evidence the hypothesis that these areas or large dimples correspond to the convolutions, as the markings would appear to be too numerous and too small. The writer prefers to regard them as an indication of some change in the actual structure of the bone due to abnormal growth, the nature of which is at present quite obscure. Leonard, discussing the Röntgen diagnosis of hydrocephaly, describes a case of hydrocephaly in which the inner table of the skull showed depressed areas, visible by ordinary light, exactly corresponding to the convolutions. He says it is only in the late form of hydrocephaly occurring after the sutures have become ossified that the pathological change takes place in the inner table. Such a condition cannot be common in oxycephaly, seeing that in the large number of skulls examined by different writers irregularity or unevenness of the inner aspect of the vault is hardly mentioned.

The skiagrams show that the frontal sinuses and antra are either absent or so rudimentary that they can barely be distinguished. The bones forming the orbital, nasal, and superior maxillary regions present an extraordinarily transparent flocculent appearance. The superior maxilla in the skiagram of Case I is greatly shortened and the last molar can be seen lying above, unerupted. The lower jaw is underhung, and is shorter and squarer than normal. The bone of the inferior maxilla appears to be of fairly normal density.

The sella turcica presents the most striking appearance; it stands out with unusual clearness and seems to be considerably enlarged and displaced backwards. The bones forming the vault show some reduction in thickness, especially in the region of the bregma, where there is often a distinct bulging. This is well seen in the skiagrams of Cases I and VI. In both there is obvious localized bulging at the vertex. For the purposes of comparison a skiagram of a normal adult skull is given (Plate 38, Fig. 23).

Other associated defects. Reference has been made to an abnormality of the elbow-joints in two of the appended cases. This defect was present in one of my cases (Case I) in which there was a deformity of the elbow-joints with inability of full extension. The same was noticed in one of Dr. Hutchison's cases (Case V). In Case II the head of the ulna appears to be thickened. This condition requires further observation as to its frequency and nature.

I must take this opportunity of thanking Dr. Walsham and Dr. Pirie of St. Bartholomew's Hospital for the excellent skiagrams they have taken of my series, and Mr. Scott of the London Hospital for those of Dr. Hutchison's cases. I am also indebted both to Professor Keith and Dr. Addison for kind suggestions on various points of anatomy.

Pathology.

The idea has long been held and is still widely prevalent that oxycephaly is essentially a congenital deformity of the head and that the chief point of interest consists in the ocular affections associated with it. Reasons will be advanced later against this point of view and in favour of the condition being regarded as a manifestation of a definite morbid process affecting more manifestly the bones of the skull than other parts of the body.

A brief review may be given here of the various hypotheses which have been advanced in explanation of the malformation of the skull. Most writers on the subject are agreed that the characteristic deformity of the skull is due to premature synostosis of certain sutures, notably the sagittal and coronal. The evidence in favour of this synostosis is very strong, as it is found in all the skulls which have been examined. The last portions of these sutures to become synostosed are those in the neighbourhood of the bregma. In some cases of oxycephaly in young subjects the anterior fontanelle has been found to be patent. As a result of the premature union of these two sutures the growth of the vault of the skull is restricted in both its antero-posterior and transverse diameters, and to accommodate the increasing bulk of the brain a compensatory increase in height takes place. Eventually the anterior fontanelle closes, but there is reason to think that this occurs at a later date than the normal, and its former site is marked by a slight protuberance with thinning of the bone, as is well shown in several of the skiagrams. The rapidly growing brain is partly accommodated by the compensatory increase in the vertical diameter, but in addition important changes are brought about at the base. The cranial fossae, especially the middle one, become deepened and thinned out by the pressure exerted by the brain. The thinnest bones naturally are most affected, and thus the orbits become deformed and shallow, the greater wings of the sphenoid in particular being pushed forward by the sphenoidal lobes, so that, as Weiss and Brugger describe it, they form the posterior instead of the lateral walls of the orbits. The temporal region is pushed outwards so that the temporal fossae become shallow or obliterated. The malformation of the vault, the cranial fossae, and the orbits may be accounted for in this way: the other bony changes will be discussed later.

What is the cause of this premature synostosis? Virchow and v. Hirschberg and others regarded the primary cause as some form of meningitis, but the evidence in favour of it is very slight. Some thickening of the pia arachnoid has been met with in one case. A diffuse meningitis would have been expected to give rise to hydrocephaly or to microcephaly with mental defect. The absence of any mental defect in the great majority of cases is in the writer's opinion a strong argument against the occurrence of meningitis of any known type.

Hydrocephaly has been advanced as a cause of the synostosis. Meltzer suggested that it was due to a reaction process in rachitic bones to the pressure

of hydrocephaly, and that after synostosis has taken place resorption of fluid occurs. Potts, in a recent paper (1910), does not accept the view that premature synostosis is a sufficient explanation of the deformity, and ascribes it to a special form of hydrocephaly, but without giving, in the writer's opinion, any evidence of its existence. A study of the recorded cases leads one to exclude rickets at any rate as an important factor in the disease. It appears to the writer that there are also cogent reasons for excluding hydrocephaly as a cause of oxycephaly. The shape of the head, the unimpaired intelligence, the absence of paralyses, the fact that such sutures or fontanelles that have been observed to be patent in the younger cases are not described as bulging, are strong arguments against this view. Further, the cerebro-spinal fluid has been examined in a few cases of oxycephaly (Behr), and was found to be normal, and the pressure was not increased. To investigate further the possible relation of hydrocephaly to oxycephaly, Dr. Walsham kindly took for me a skiagram of the head of an adult hydrocephalic patient, aged 52, for comparison with that of the adult oxycephalic cases. The bones of the skull presented a totally different appearance; they were uniformly homogeneous and did not show the 'digital markings' met with in oxycephaly, and further, even at this age, the sutures were still ununited. The same may be said of the skull of a hydrocephalic child I have recently examined.

What is the cause of this premature synostosis if it is not due to some form of meningitis, a rachitic condition of the bones, or hydrocephalic pressure? In the writer's opinion it depends upon some deeper underlying cause, the nature of which at present remains obscure, and in his opinion the term 'oxycephaly' is a misleading one, as it implies as the essential condition deformity of the head. It seems probable that the condition should be looked upon from a wider point of view, the changes in the bones of the skull being only one of its manifestations and the most obvious. Bertolotti has recently made a valuable contribution to our knowledge of the subject; in this he regards oxycephaly as 'un syndrome osseux qui peut être bien plus étendu que ne voudrait signifier son nom, et qui correspond, non pas seulement à des altérations de la calotte, mais aussi de la base du crâne, des os de la face et enfin de plusieurs autres parties du squelette'.

With this the writer is in agreement, although he does not agree with Bertolotti that oxycephaly is a dystrophy of rachitic origin. It is a noteworthy point that nearly all those who have investigated the skiagraphical appearance of the skull have noted the extraordinary changes which are present in the sella turcica, and it may possibly be shown that the alterations in the structure and density of the bone and premature synostosis may be the result of some change or abnormal development in the pituitary body. This must of course be regarded as a mere conjecture, for no post-mortem observations have been made with regard to it.

Professor Keith's recent lectures on 'Prehistoric Man' and the suggestions he has made as to the possible part played by the pituitary body in the

development of racial characteristics may be brought forward in this connexion. The skull is not the only part involved in the condition of oxycephaly; other parts are also affected. There are many points which require investigation, notably the histological features of the affected bones and the alterations and changes in the joints such as were present in Case I.

Cause of the optic atrophy. A brief summary must be given here of the various explanations of the optic atrophy which is almost invariably present. Virchow regarded it as secondary to a neuritis due to some form of meningitis; Ponfick and others, that it is due to papillary stasis caused by narrowing of the optic foramen. Friedenwald considered that the papillitis and atrophy are the result of direct pressure exerted, owing to the synostosis, by the growing brain comparable to that in cerebral tumour. This view is also held by Dorfmann, who points out as a result of examination of skiagrams, that, as the changes at the base of the skull advance, the optic chiasma becomes unduly exposed to pressure.

Behr suggests that intracranial pressure provokes a stasis of the papilla, and that the peripheral fibres of the optic nerve undergo atrophy, leading to concentric diminution of the visual field. Atrophy of these peripheral fibres takes place with resulting diminution in size of the nerve and relief of pressure. In this way he explains the comparative rarity of complete blindness.

The writer inclines to the view that the optic atrophy is the direct result of pressure exerted by the growing brain. The cases of oxycephaly in which papillitis or optic atrophy is absent are explained by the compensatory expansion of the skull in abnormal directions being adequate for the increasing growth of the brain.

Treatment.

So far as is known there is no treatment for the cause of oxycephaly. Headache, which is such a frequent symptom, was relieved in two of my cases by the administration of potassium iodide. Operative treatment has been employed with a view to save the sight. One of Dorfmann's cases, a girl aged 4, had well-marked papillitis and suffered from severe headaches and insomnia. In 1907 Eiselsberg trephined her. The bone was found to be then 1½ mm. in thickness, the dura mater normal, the veins distended. Three weeks later the swelling of the disks had diminished 1 diopter, and the headache and sleeplessness were relieved. Trephining to relieve pressure would seem a reasonable method of treating cases of oxycephaly in children when papillitis without atrophic changes is present, as probably all such cases subsequently become practically if not completely blind.

Before proceeding to the detailed account of the series of seven cases, I must express my gratitude to Dr. Robert Hutchison for permitting me to include his three cases in this series, and for the trouble he has taken in procuring the photographs and skiagrams which illustrate them.

Case I. A. R., male, 23, storekeeper: photographs (Plate 30), skiagram of skull (Plate 31). Did not know at what age the exophthalmos or head deformity began. Eyesight defective as long as he could remember, and always worse in left eye. Never went to school, owing to bad eyesight, but taught himself to read and write with his father's help. Did not suffer from headache and had never had fits. His mother (whom I have seen) had exophthalmos and malformation of the superior maxilla, but the vault of the cranium was not definitely oxycephalic. Parents English.

Head typically oxycephalic (photographs, Plate 30). The bregma was prominent. Extreme proptosis; the right eye was more prominent than the left and occasionally became dislocated forwards in front of the eyelids. Divergent squint and nystagmus, could distinguish only moving objects with left eye, can read print with right eye. Mr. Holmes Spicer reports: 'Right pupil unusually small, but reacts normally. Left pupil larger than right and insensitive to light. Both optic disks very white; the *right* disk has ill-defined edges and a definite white fibrosis-looking prolongation up and inwards suggesting a previous neuritis; the lamina cribrosa also is not visible, but the vessels are quite clear. The *left* disk is quite clear and the vessels well defined—the lamina cribrosa is not seen. The condition of the disks is quite compatible with previous neuritis. Right visual field much reduced.'

The nose was strongly deflected to the right; this was stated to be the result of an injury. The superior maxilla was very deformed, it was shortened from before backwards. The hard palate formed a narrow, very pointed arch. The second and third molars were absent in the upper jaw and could be made out in the skiagram lying above in what appeared to be the rudimentary antra. Lower jaw underhung, teeth normal in number. Sense of smell lost. Hearing and taste normal.

Elbows could not be fully extended (Plate 30, Fig. 3, and Plate 31, Figs. 5 and 6). Internal condyles unduly prominent, some creaking in the joints: the head of both radius and ulna appears thickened. He could not raise the arms to the horizontal; obvious creaking in both shoulder-joints, but no change found in the bones. No other malformations present.

Skiagram (Plate 31, Fig. 4) of the skull shows very distinctly the bulging and thinning of the bone in the bregmatic region (?) and the digital markings over the whole of the vault. The most striking abnormality is the altered shape and depth of the middle fossa, and the pushing forward of the posterior wall of the orbit. The sella turcica is pushed backwards and is deepened (cf. skiagram of normal adult skull, Plate 38, Fig. 23).

Dr. R. J. Gladstone very kindly took the following measurements of this case two years ago:—

Height, 5 ft. $2\frac{3}{4}$ in. (1522 mm.).

Circumference of head, 506 mm. Longitudinal arc, 345 mm. Transverse arc, 352 mm.

Diameters of head: L. 179 mm. B. 138 mm. H. 143 mm. Mi. F. 106 mm.

'Index of size' of head, 3532. (The average 'index of size' of males, 5 ft. 3 in. height, is 3850.)

Cephalic breadth index, 76.

Cephalic height index, 79·8.

Horizontal arc, measured from the centre of the external auditory meatus round the sub-nasal point to the centre of the meatus of the opposite side = 231 mm. (Average about 255 mm.)

Average diameters of head in fifty male subjects, aged 20 to 46, measured in the post-mortem room, Middlesex Hospital:—

Length: glabella to occipital point	190·8 mm.
Breadth: greatest transverse diameter . . .	149·5 mm.
Height: bi-auricular line to vertex	134·8 mm.

There is thus considerable diminution in the length and breadth, and increase in height.

	Normal	Case A. R.
Average cephalic breadth index . . .	77·6	76·0
„ „ height „ . . .	70·0	79·8

Case II. E. T., female, 44, charwoman: photographs and skiagram of skull (Plate 32). Did not know when the deformity of the head began. Blind in left eye as long as she could remember. Had suffered with headache, usually vertical, all her life. Single. No past illnesses of importance. One of twins, the other having died in infancy. English descent.

Typically oxycephalic: dark sallow complexion, greasy skin. Nose deflected to left. Hard palate asymmetrical. Distinct exophthalmos; divergent squint; some nystagmus; blind in left eye; pupils react normally. Mr. Holmes Spicer reports on the eyes: 'Some fibrous vitreous opacities; both optic disks very white, having a convex or heaped-up appearance and with a somewhat uneven edge in the left eye. Marked whitening of the veins in both eyes extending far out from the disk. In the central region of both is a large number of small deposits in the choroid, smaller in the right than the left (? hyaline bodies). Visual field of right eye greatly diminished.'

Sense of smell defective. Taste and hearing good. No malformation of the limbs or extremities.

Case III. J. P., male, 15: photographs and skiagram (Plate 33). Hebrew. Up to 4 years of age was like other children, according to his mother's account, and the eyes and head were natural. Then the sight began to fail and the head gradually altered its shape and the eyes became prominent. Never fits —suffered from frequent headache. Past and family history good. Very intelligent, learned blind reading at an early age. Typical oxycephaly with exophthalmos, divergent squint, and marked nystagmus. Could barely distinguish large objects. Slight deflexion of nasal septum. Hard palate arched but not nearly so deformed as in Case I. Hearing, taste, and smell normal. There were no other malformations. The skiagram shows the same features as in Case I, the distinct digital markings and the changes in the orbits and the base.

Case IV. J. R., male, 14: photographs and skiagram (Plate 34, and Plate 35, Fig. 16). Eyes large at birth and head noticed to be peculiarly shaped. Suffered from frequent occipital headaches. Had never had fits. Memory not good and was only in fourth standard. Was the first and only child, difficult labour, breech presentation. Family history good.

Oxycephalic skull. Exophthalmos and divergent squint. No nystagmus. Mouth-breather. Nasal septum deflected. Superior maxilla shortened. Vision good. No optic atrophy. Ear poorly developed, tragus rudimentary. Sense of smell, taste, and hearing normal.

Peculiar malformation of thumbs; the first phalanx was curved outwards towards the radial side, the outer or radial side being shorter than the inner.

The skiagram of the skull shows very distinctly the characteristic changes at the base and the digital markings.

Case V (Dr. Hutchison's case). H. F., female, 5. (Plate 35, Fig. 17.) Parents English. Youngest of seven, others normal; two died in infancy. Three miscarriages, one a monster with 'head like a cow'. Full term child. Was born with prominent eyes and misshapen head. Previous health good. Never fits. Oxycephalic head with flat occiput, prominent bregmatic region. Exophthalmos, divergent squint, defective vision, optic disks atrophic. Mouth-breather; palate highly arched; superior maxilla shortened from before backwards. Elbows could not be fully extended as in Case I; internal condyle prominent. The skiagram

showed thickening of the heads of both radius and ulna. Thumbs shortened; first phalanx curved outwards as in Case IV; only two phalanges in left index finger, one long and a short terminal. Toes webbed and great toes very short.

Intelligence good. Taste and hearing good: sense of smell appears to be defective.

Skiagram of head not obtained.

Case VI (Dr. Hutchison's case). S. F., female, 12. (Plate 36.) Ninth of ten children. Rather difficult full term labour. Mother had a severe fright in the sixth week of pregnancy. Jewish parents. Deformity of head and prominent eyes noticed at birth, could see till three years old.

Oxycephalic head. Exophthalmos with divergent squint. Constant, irregular, chiefly lateral movements of the eyes. Pupils react sluggishly to light. One of the eyeballs became dislocated forwards on one occasion. Disks show well-marked atrophy of secondary type with white lines along course of vessels: cannot see light. Sense of smell lost, taste good, hearing somewhat impaired. Suffers from severe headaches. Is said to be 'very clever'. Adenoid facies; high narrow palate; tubercle of Atlas very prominent; thumbs very broad; little fingers kinked like those of a Mongol; viscera normal.

Skiagram of skull (Plate 36, Fig. 20). Bulging in bregmatic region. Digital impressions very well marked.

Case VII (Dr. Hutchison's case). M. W., male, 9. (Plate 37.) Said to have been normal at birth. At 18 months had convulsions while teething, which recurred at intervals for 3 months. Had a fall and whooping-cough about this time, when sight was first noticed to fail. Born in Warsaw. The boy is clever, can find his way anywhere, and is distinctly musical. Mother had three miscarriages. Marked oxycephaly: bregmatic region pointed. Circumference of skull, 19 inches; coronal suture, 13 inches. Vision very defective. Optic atrophy. Can distinguish light and dark, but cannot see fingers. Hands, feet, and viscera normal.

Skiagram shows very strongly marked 'digital impressions'. The vault is thin in the bregmatic region.

REFERENCES.

1. Algyogyi, H., *Wiener med. Wochenschr.*, 1908, lviii. 1942.
2. Atgier, *Bull. et Mém. Soc. d'anthrop. de Paris*, 1901, 5e sér., ii. 95.
3. Beaumont, W. M., *Trans. Ophthal. Soc. U. K.*, 1910, xxx. 44.
4. Behr, *Archives d'Ophthal.*, 1910, xxx. 714.
5. Behr, *Ann. d'Oculistique*, Paris, 1910, cxliv. 217.
6. Bertolotti, *Presse médicale*, Paris, 1910, ci. 946.
7. Bourneville et Boncour, *Bull. et Mém. Soc. d'Anthrop. de Paris*, 1902, 5e sér., iii. 35.
8. Carpenter, G., *Proc. Roy. Soc. Med.*, 1908–9, ii, 'Sect. Dis. of Children,' 45.
9. Carpenter, G., *Trans. Ophthal. Soc. U. K.*, 1909, xxix. 157.
10. Carpenter, G., *Reports Soc. Study of Dis. in Children*, Lond., 1901, i. 110.
11. Coats, G., *Trans. Ophthal. Soc. U. K.*, 1907, xxvii. 211.
12. Cohen, C., *Klin. Monatsbl. für Augenheilk.*, 1906, N. F., ii. 517.
13. Dodd and McMullen, *Lancet*, Lond., 1903, ii. 1665.
14. Donaldson, E., *Trans. Ophthal. Soc. U. K.*, 1903, xxiii. 264.
15. Dorfmann, R., *Archiv für Ophthal.*, Leipz., 1908, lxviii. 412.
16. Dorrell, E. A., *Trans. Ophthal. Soc. U. K.*, 1910, xxx. 158.
17. Enslin, *Archiv für Ophthal.*, Leipz., 1904, lviii. 151.
18. Enslin, *Ophthalmoscope*, 1904, ii. 233.
19. Fletcher, H. M., *Proc. Roy. Soc. Med.*, 1908–9, ii, i. Clin. Sect., 113.
20. Ford, R., *Ophthalmoscope*, 1907, v. 199.

21. Friedenwald, H., *Amer. Journ. Med. Sci.*, 1893, N. S. cv. 529.
22. Friedenwald, H., *Archives of Ophthal.*, N. York, 1901, xxx. 405.
23. Gräfe, A., *Archiv für Ophthal.*, Berlin, 1866, xii, Abth. ii. 114.
24. Groenouw, A., *Gräfe-Sämisch's Handb. der Augenheilk.*, 2. Aufl., 1904, xi, Abth. i. 257.
25. Hanotte, M., *Thèse de Paris*, Paris, 1898.
26. Harman, N. B., *Trans. Ophthal. Soc. U. K.*, 1910, xxx. 106.
27. v. Hirschberg, J., *Centralbl. für Augenheilk.*, 1883, 1.
28. v. Hirschberg, J., und Grunmach, E., *Berl. klin. Wochenschr.*, 1909, xlvi. 191.
29. Hutchison, R., *Proc. Roy. Soc. Med.*, 1909–10, iii, 'Sect. Study Dis. Children,' 125.
30. Krauss, W., *Zeitschr. für Augenheilk.*, 1907, xvii. 432.
31. Krauss, W., *Ophthalmoscope*, 1908, vi. 530.
32. Leonard, *Archives of the Röntgen Ray*, 1911, xv. 336.
33. Manz, *Bericht über die* 19. *ophthal. Versamml.*, Heidelberg, 1887, 18.
34. Meltzer, *Neurol. Centralbl.*, Leipz., 1908, xxvii. 562.
35. Merle, P., *Nouv. Iconog. de la Salpétrière*, Paris, 1908, xxi. 349.
36. Michel, J., *Archiv für Heilkunde*, 1873, xiv. 39.
37. Morax et Patry, *Ann. d'Oculistique*, Paris, 1904, cxxxii. 120.
38. Moutier, *Revue neurologique*, Paris, 1908, xvi. 1258.
39. Nettleship, E., *Trans. Ophthal. Soc. U. K.*, 1904–5, xxv. 383.
40. Nettleship, E., *Trans. Ophthal. Soc. U. K.*, 1887, vii. 222.
41. Oberwarth, E., *Archiv für Kinderheilk.*, 1905, xlii. 79.
42. Oliver, C. A., *Amer. Journ. Med. Sci.*, N. S., 1902, cxxiii. 4.
43. Oppenheim, *Lehrbuch der Nervenkrankh.*, 1905, 741.
44. Papillant, *Revue de l'École d'anthropologie de Paris*, 1903.
45. Paton, L., *Trans. Ophthal. Soc. U. K.*, 1904–5, xxv. 364.
46. Paton, L., *Trans. Ophthal. Soc. U. K.*, 1907, xxvii. 215.
47. Patry, A., *Thèse de Paris*, 1905.
48. Patry, A., *Recueil d'Ophthal.*, Paris, 1906, 3e sér., xxviii. 158.
49. Patry, A., *Ann. d'Oculistique*, Paris, 1906, cxxxv. 314.
50. Patry, A., *Recueil d'Ophthal.*, Paris, 1906, 3e sér., xxxviii. 156.
51. Ponjick, *Breslauer ärztl. Zeitschr.*, 1885, vii. 54.
52. Potts, W. A., *Reports Soc. for Study of Dis. in Children*, Lond., 1908, viii. 407.
53. Power, H., *Trans. Ophthal. Soc. U. K.*, 1894, xiv. 212.
54. Rau, *Sitzungsber. d. Berliner ophthalm. Gesellschaft*, Mai 1899.
55. Schuller, G., *Centralbl. für Augenheilk.*, 1881, v. 236.
56. Stephenson, S., *Proc. Roy. Soc. Med.*, Lond., 1908–9, ii, 'Sect. Study Dis. Children,' 229.
57. Stephenson, S., *Brit. Journ. of Children's Diseases*, 1905, ii. 491.
58. Stood, *Klin. Monatsbl. für Augenheilk.*, 1884, xxii. 248.
59. Terrien, *Brit. Journ. of Children's Diseases*, 1910, vii. 176.
60. Tucker, E. F. G., *Lancet*, Lond., 1904, ii. 88.
61. Uhthoff, W., *Klin. Monatsbl. für Augenheilk.*, 1905, xliii, i. 1.
62. Velhagen, K., *Münch. med. Wochenschr.*, 1904, li. 1389.
63. Velhagen, K., *Ophthalmoscope*, 1905, iii. 83.
64. Videcky, *Zeitschrift für Augenheilk.*, 1907, xviii.
65. Vortisch, *Inaug. Diss.*, Tübingen, 1901.
66. Vossius, A., *Klin. Monatsbl. für Augenheilk.*, 1884, xxii. 172.
67. Weiss, L., and Brugger, O., *Archives of Ophthal.*, N. York, 1895, xxiv. 55.

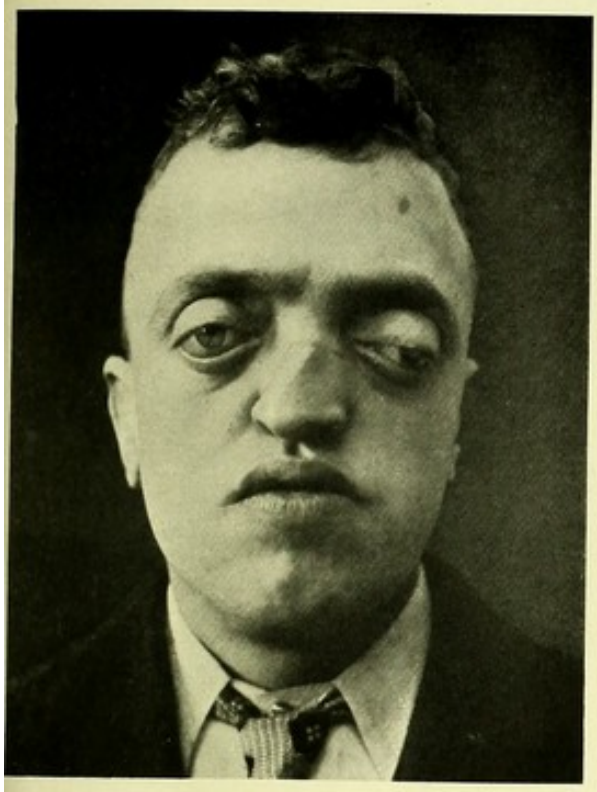

FIG. 1

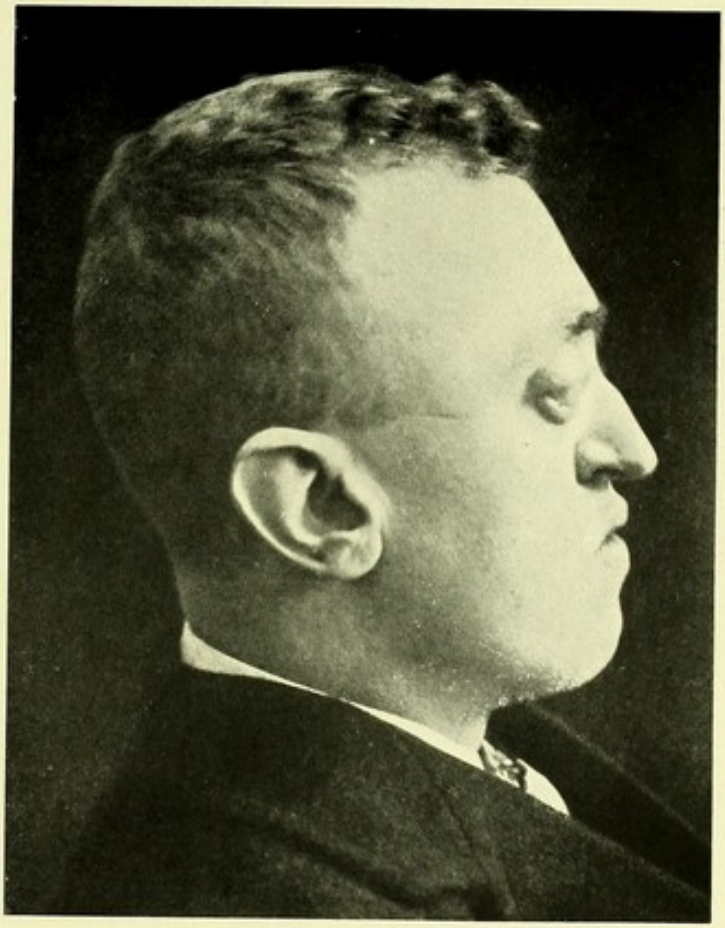

FIG. 2

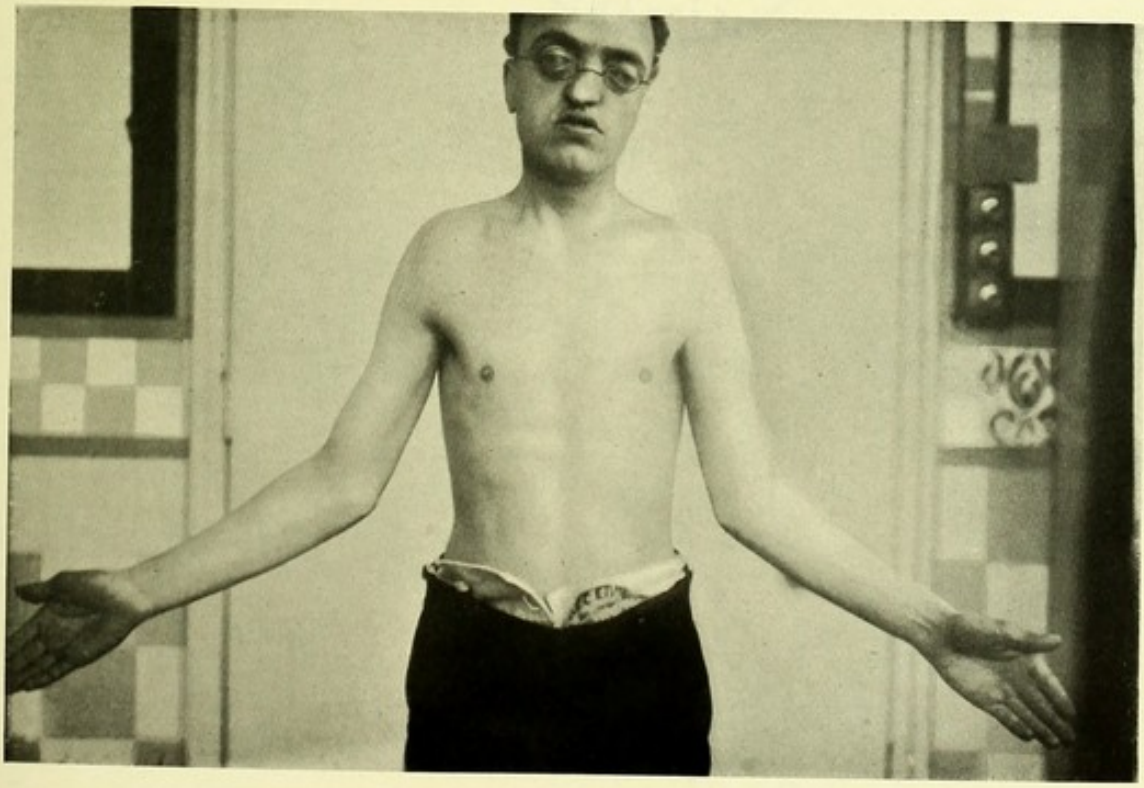

FIG. 3

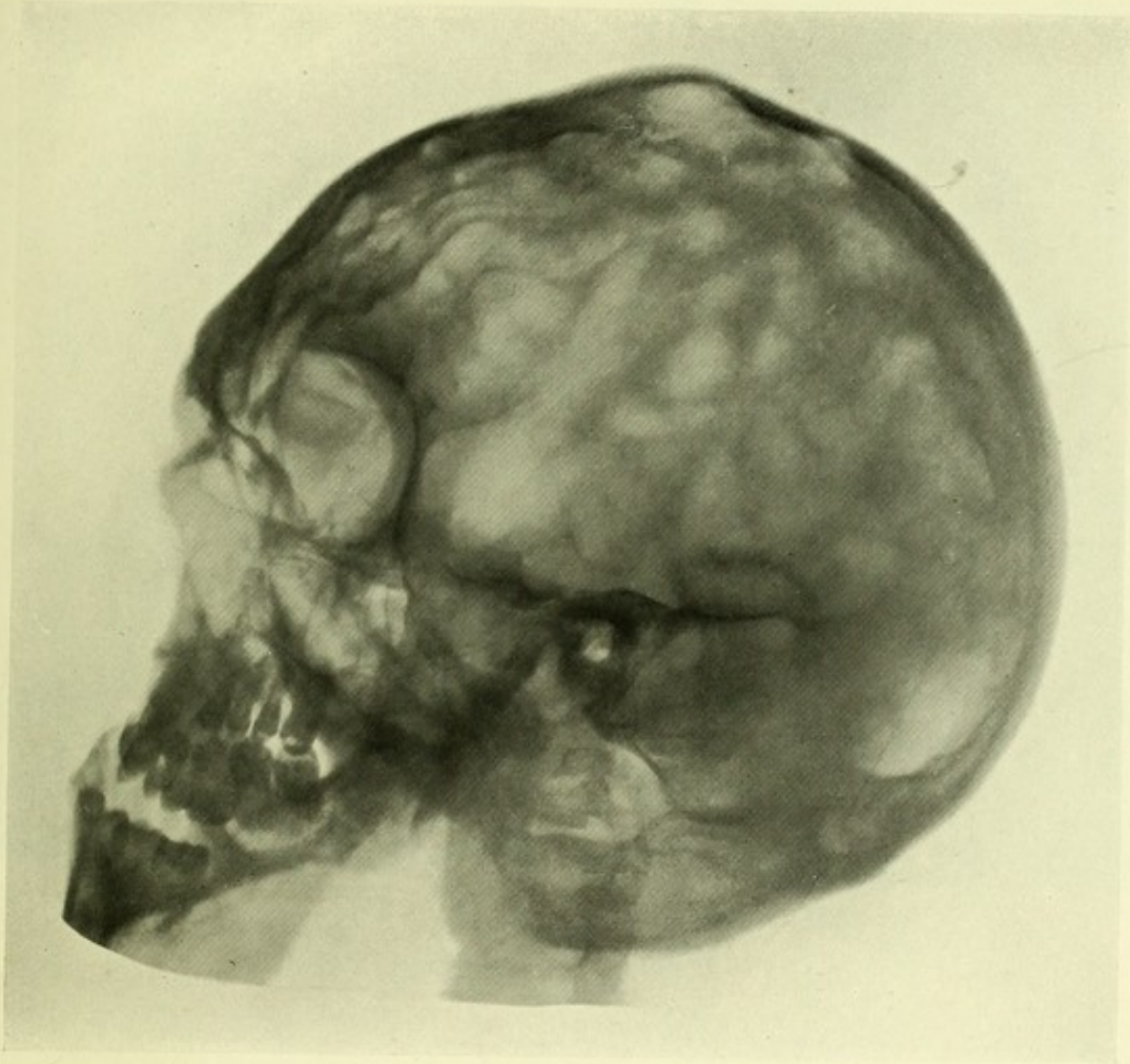

FIG. 4

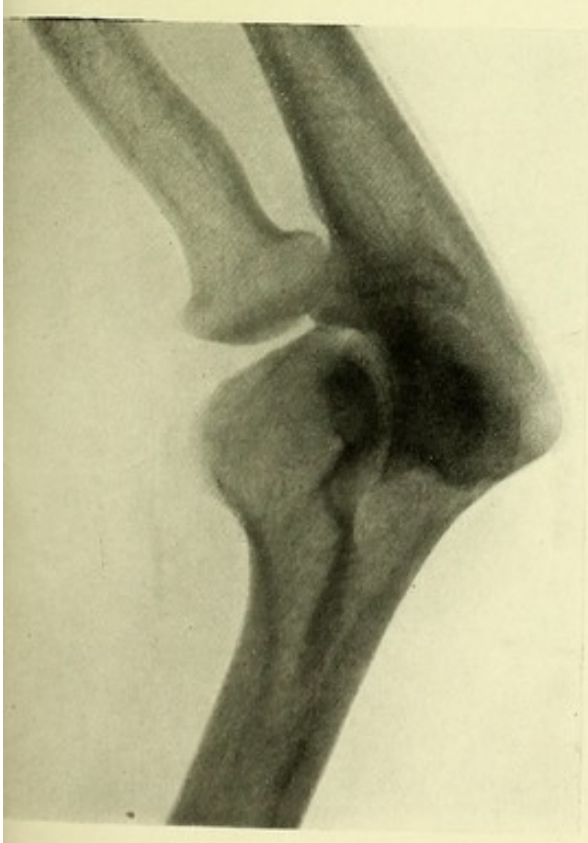

FIG. 5

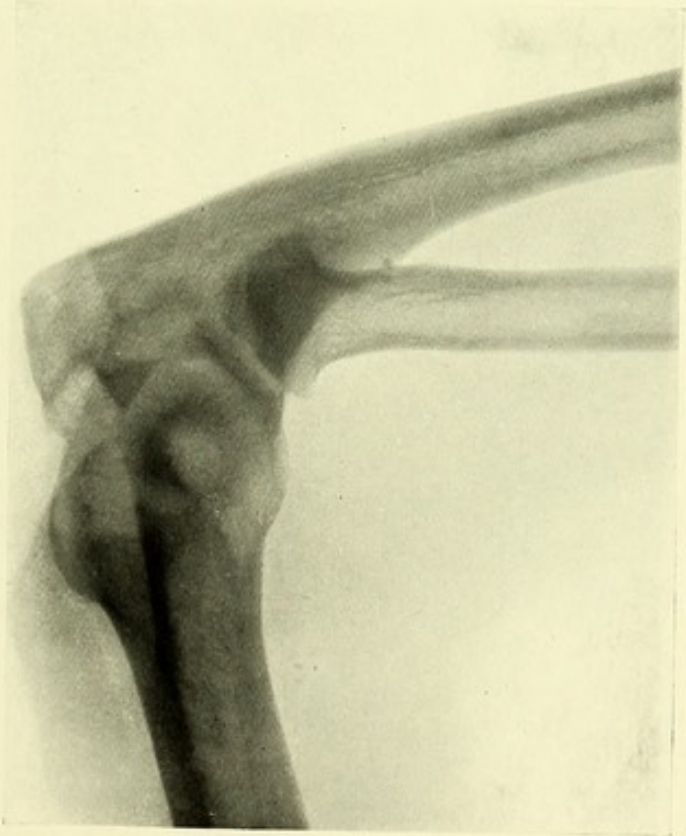

FIG. 6

ROYAL COLLEGE OF SURGEONS OF ENGLAND
28 MAY 15
LIBRARY

FIG. 7

FIG. 8

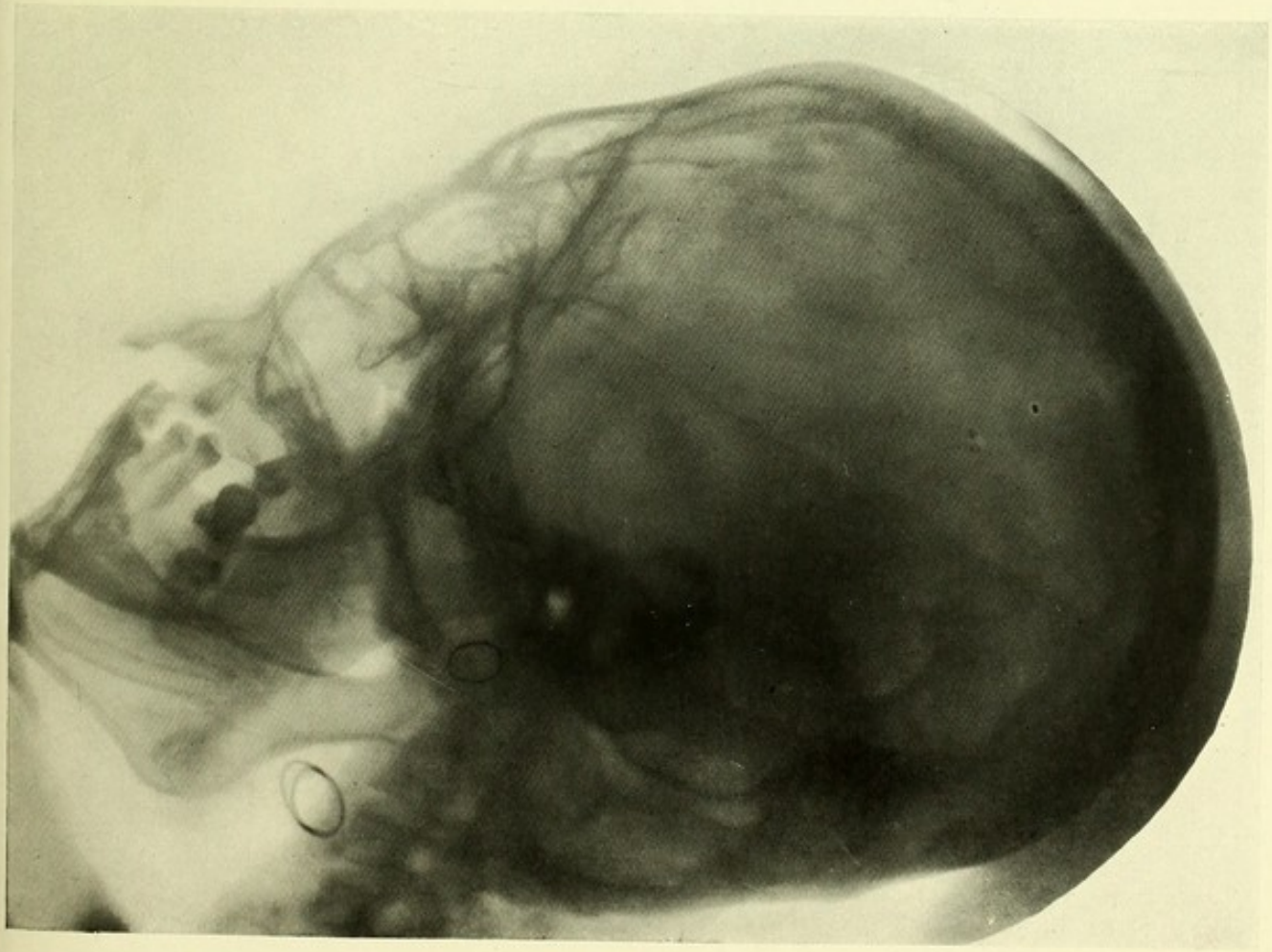

FIG. 9

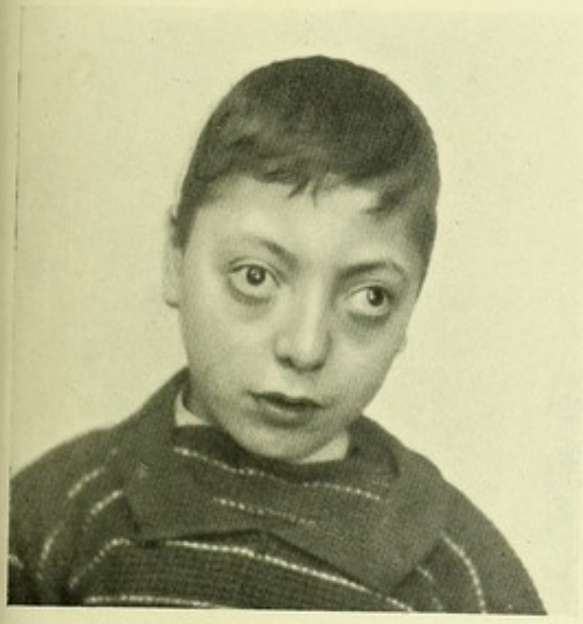

FIG. 10

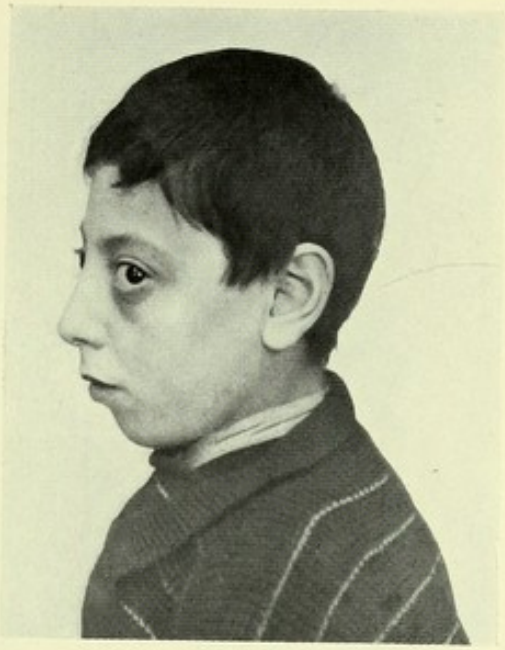

FIG. 11

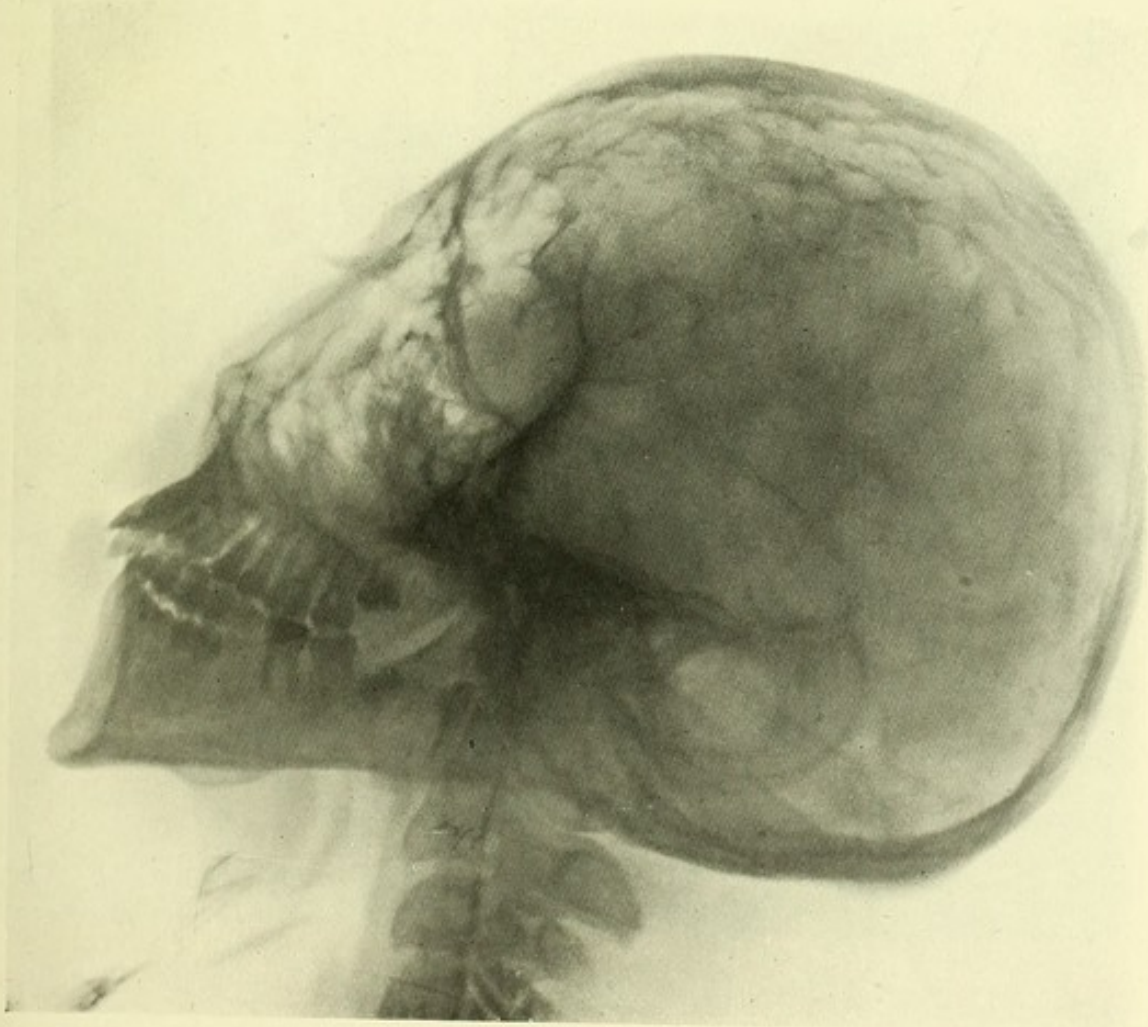

FIG. 12

ROYAL COLLEGE OF SURGEONS OF ENGLAND
28 MAY 15
LIBRARY

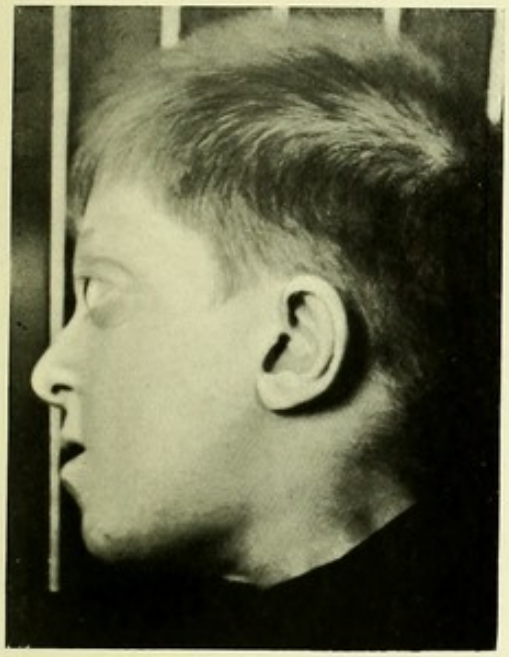

FIG. 13

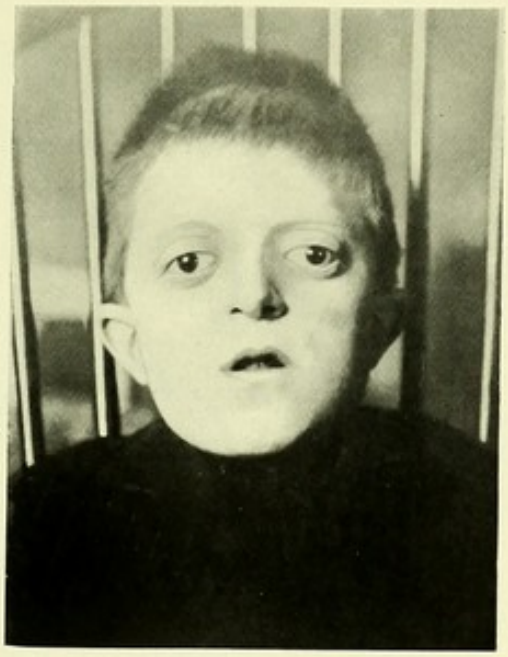

FIG. 14

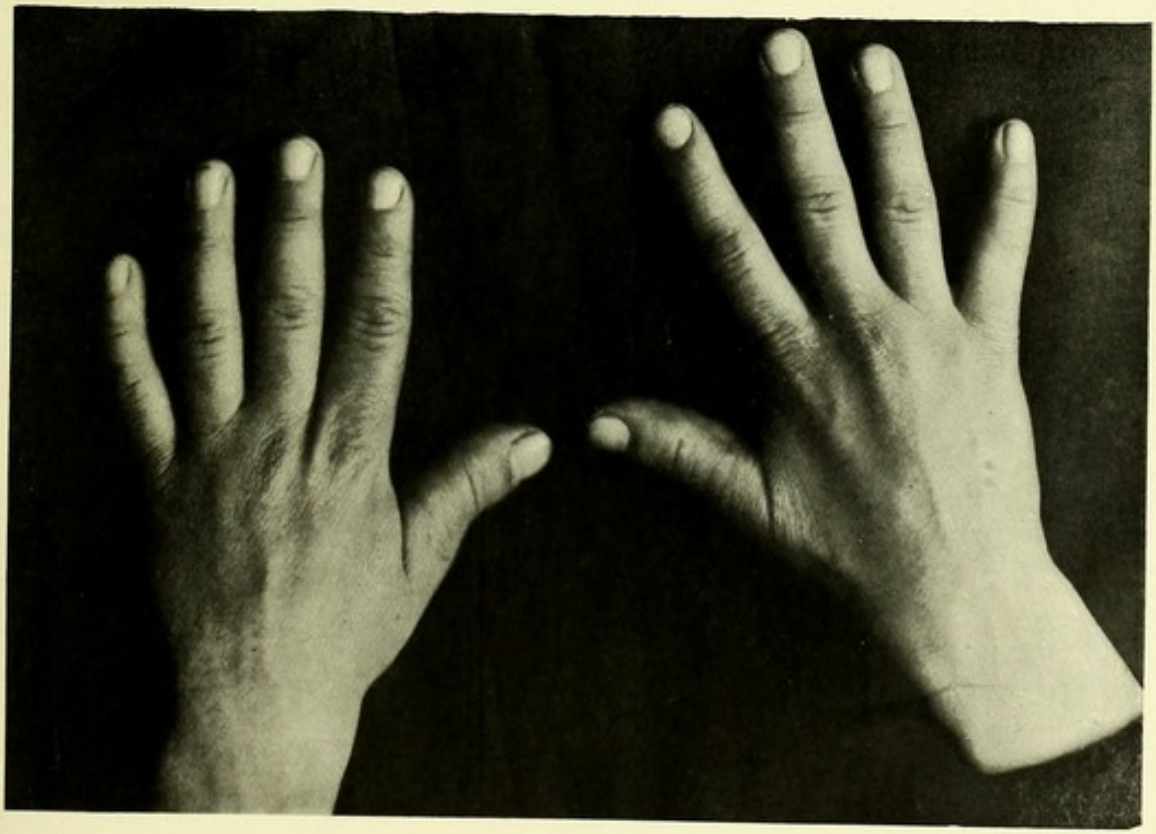

FIG. 15

ROYAL COLLEGE OF SURGEONS OF ENGLAND
29 MAY 15
LIBRARY

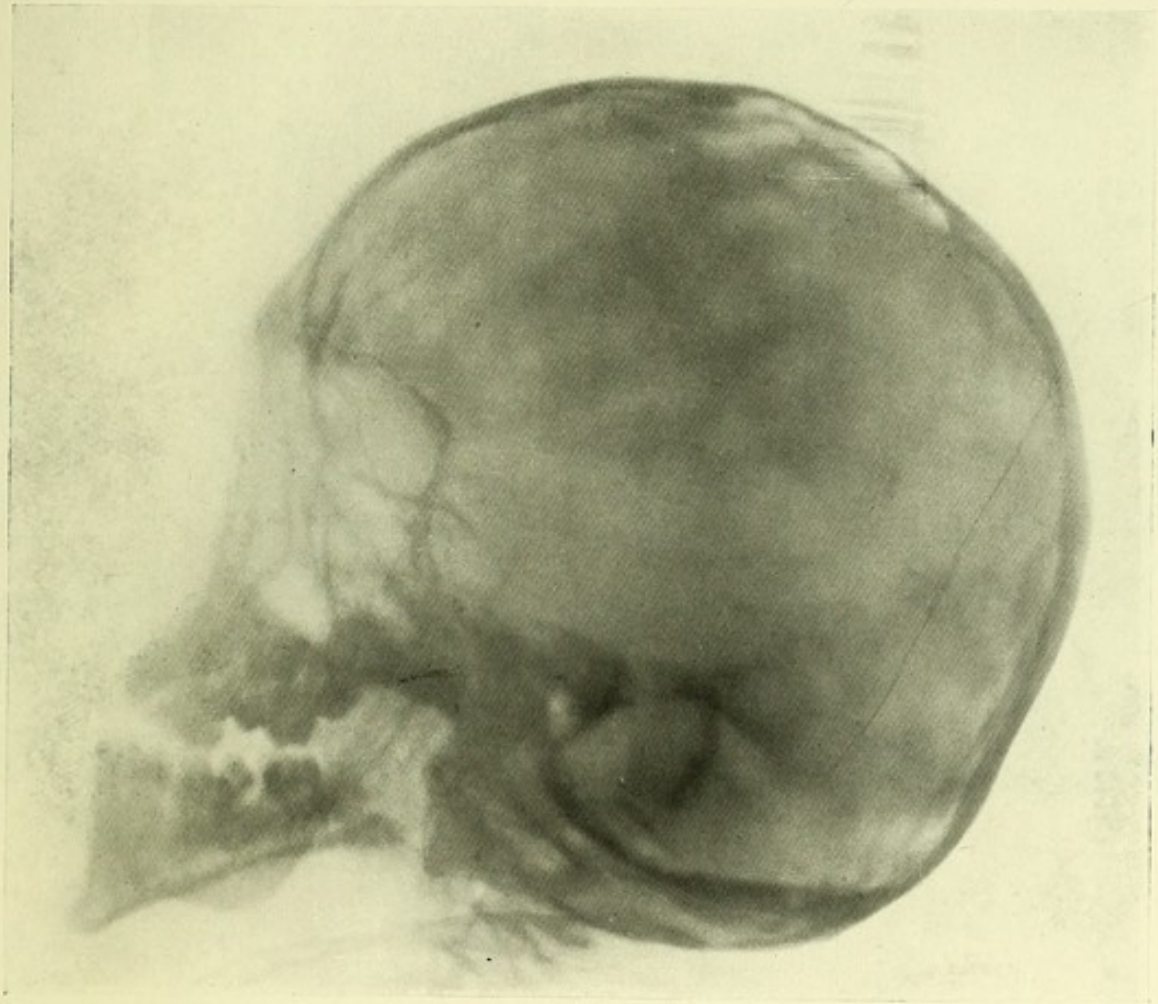

FIG. 16

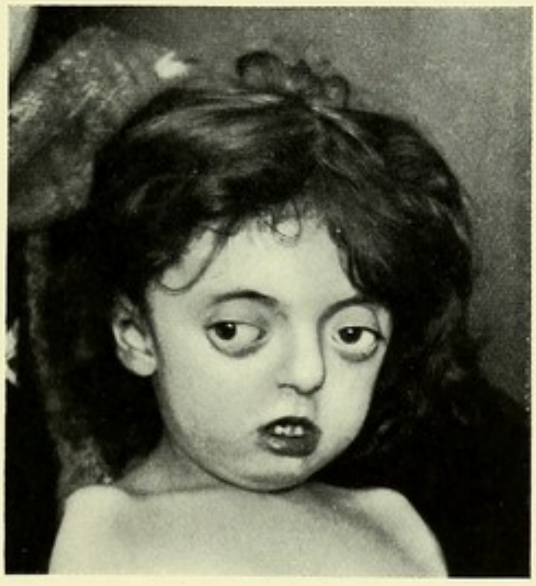

FIG. 17

FIG. 18

FIG. 19

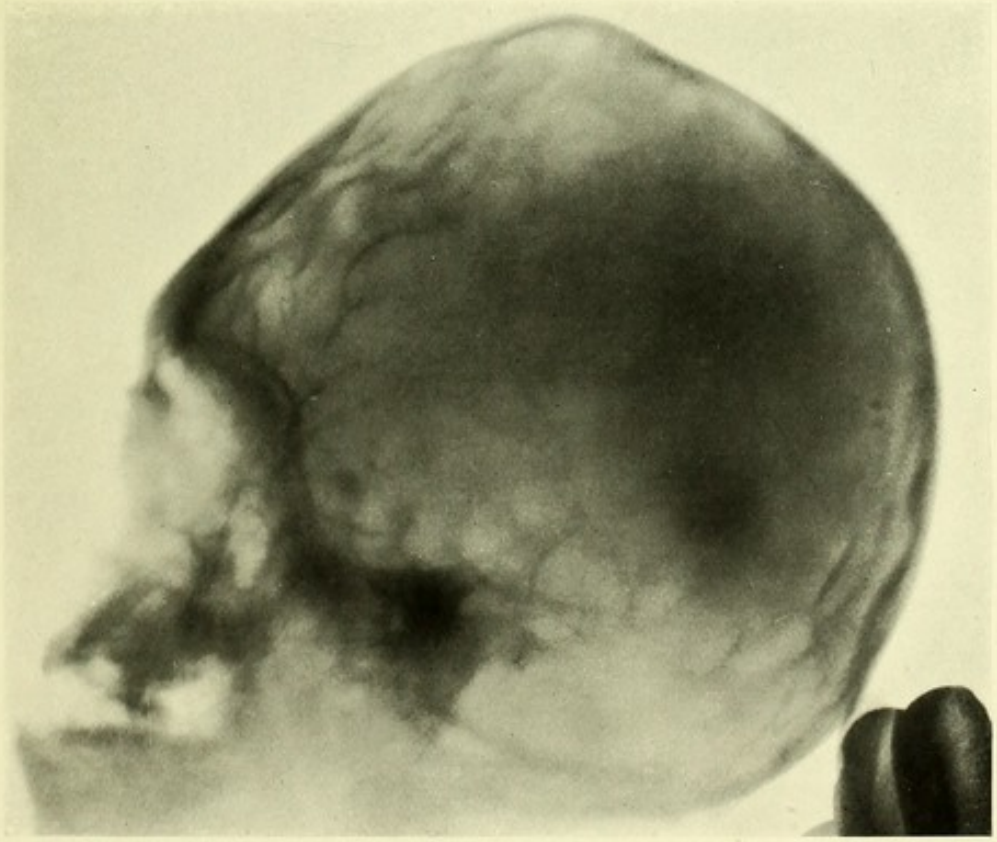

FIG. 20

ROYAL COLLEGE OF SURGEONS OF ENGLAND
LIBRARY

ROYAL COLLEGE OF SURGEONS OF ENGLAND
LIBRARY

FIG. 21

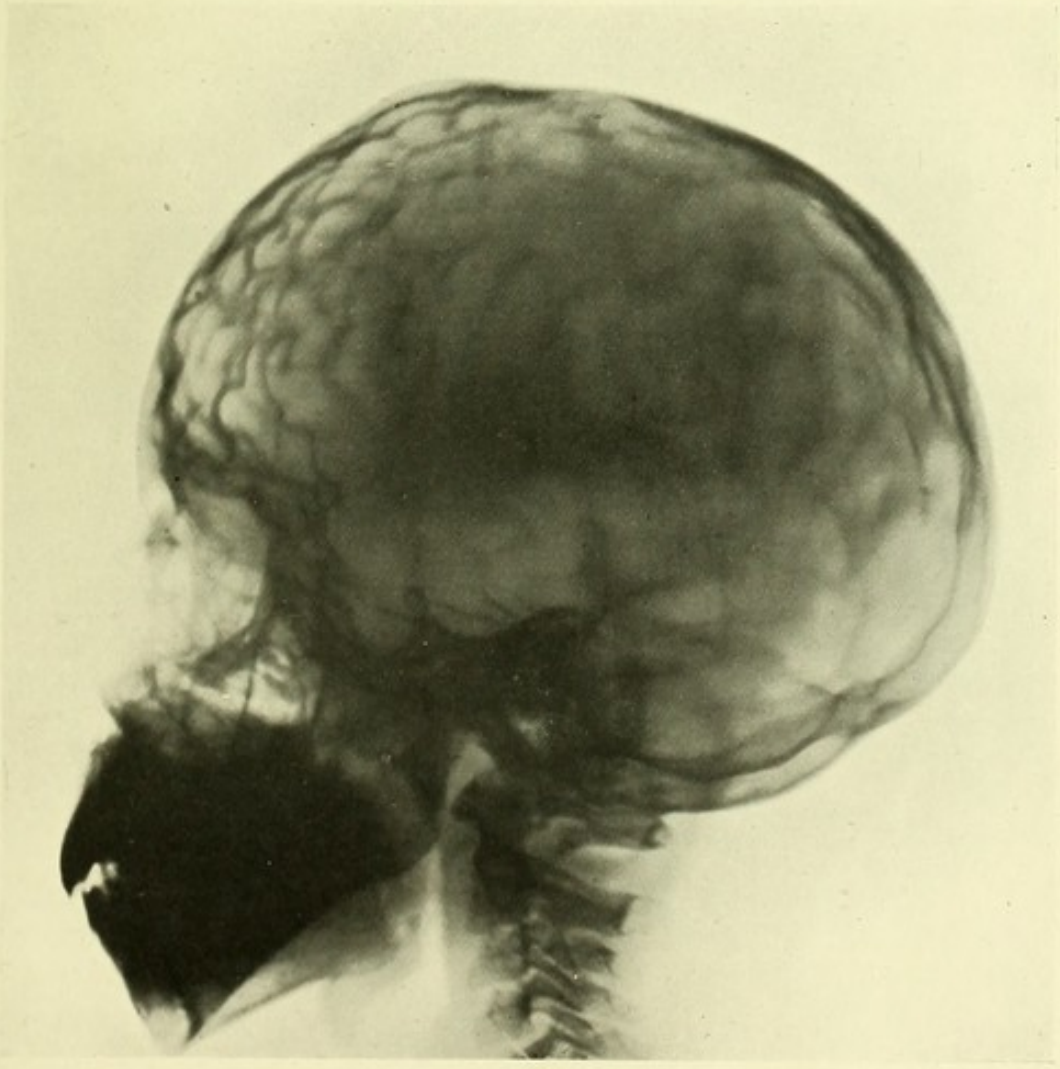

FIG. 22

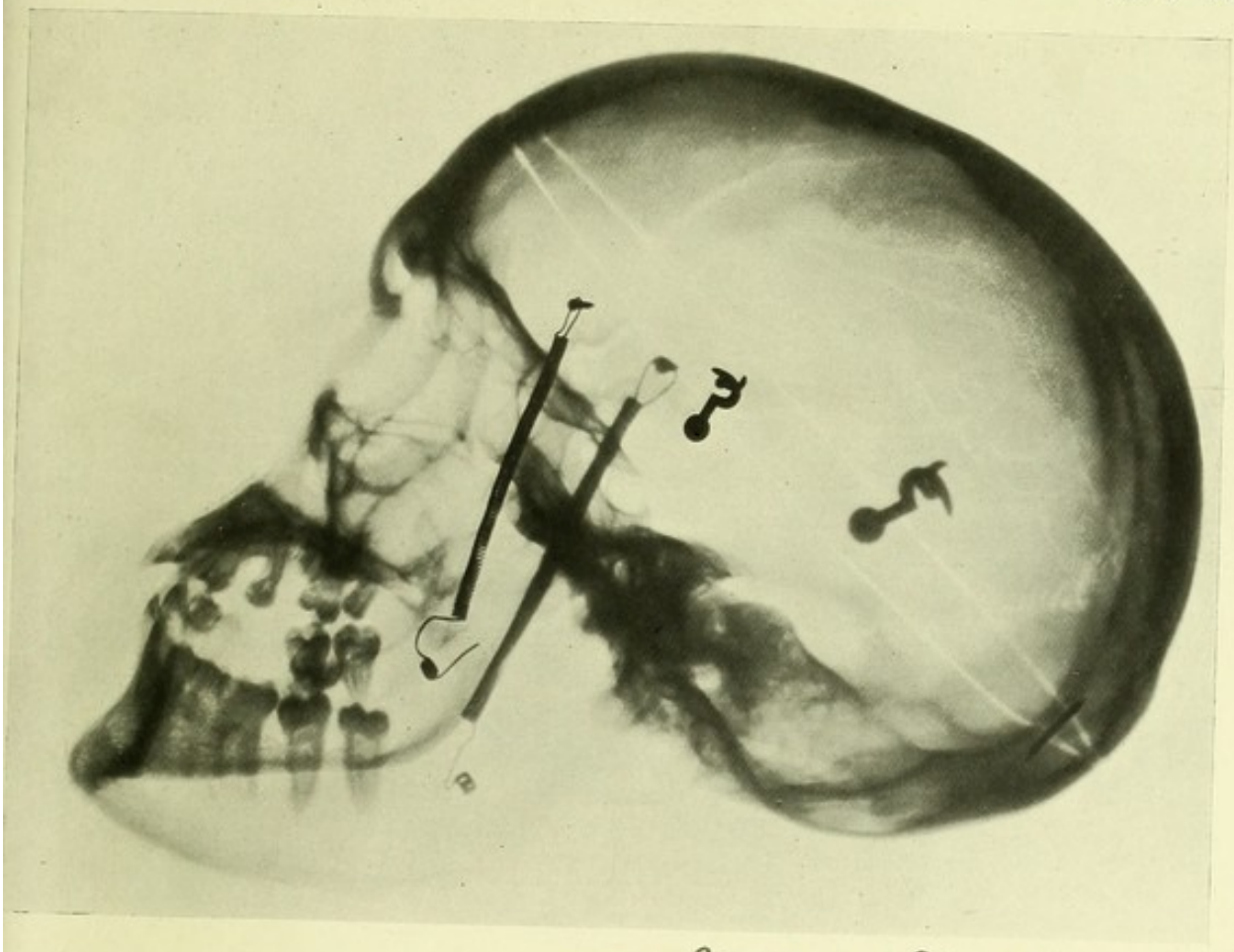

FIG. 23 Normal Skull.

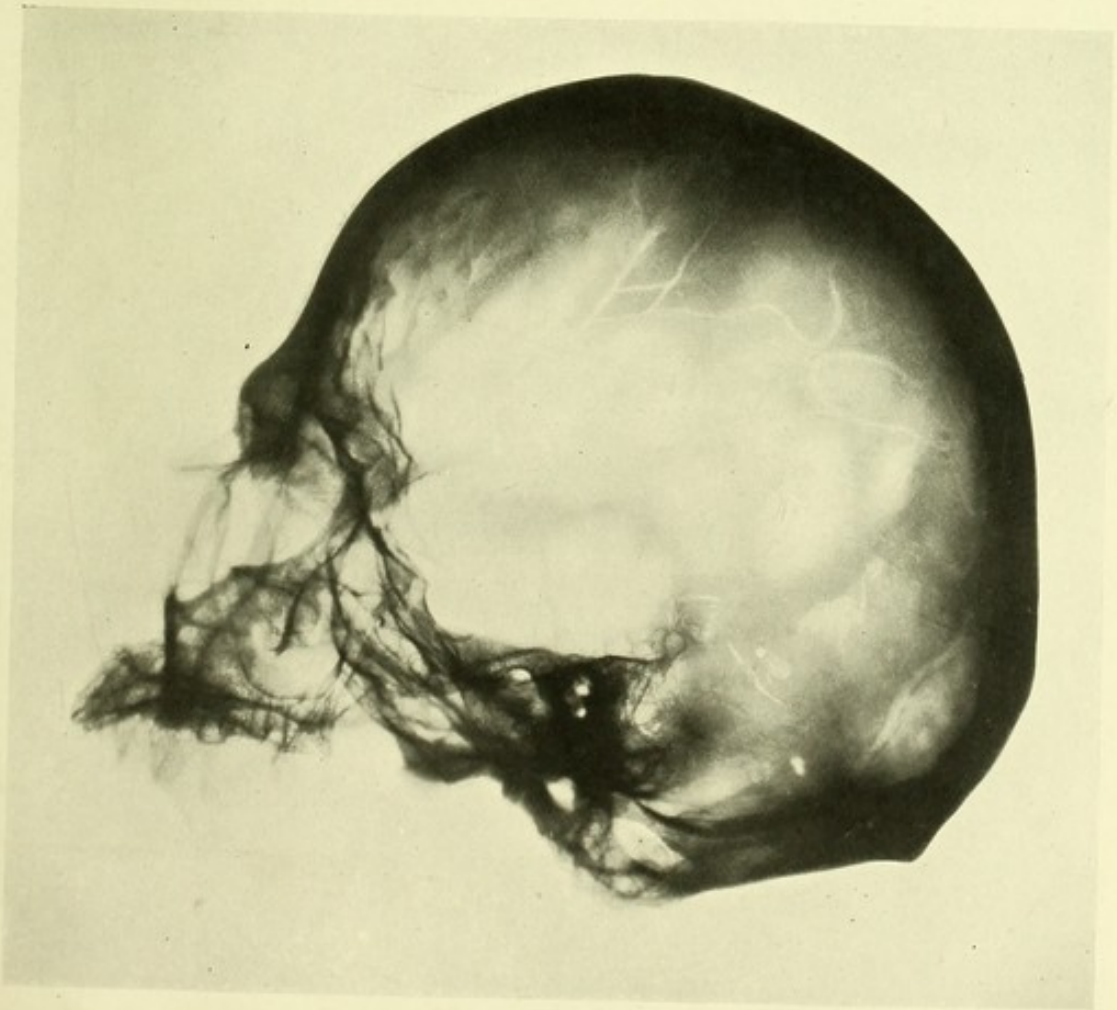

FIG. 24 Oxycephalic Skull
R. Coll. Surgeons.