CONGENITAL PERFORATIONS OF THE SKULL IN
RELATION TO THE PARIETAL BONE

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CONGENITAL perforations of the skull
are uncommon lesions and most
occur in the parietal bone, when they
are called parietal fenestrae or enlarged
parietal foramina. Any other major defects
are rare and are not well documented. The
management of the patient with these lesions
is far from clear. Two cases are described in
which the gaps were of such size as to make
cranioplasty desirable.

CASE HISTORIES

Case 1. G.M.O., a girl aged 4½ years, was ad-
mitted to Dunedin Hospital on Nov. 3, 1960. She
had been seen first on Aug. 1, 1956 shortly after
birth when it had been noticed that there was a
gap in the middle of the back of the skull. This
was triangular in shape and measured about 3.5
cm. both across the base and in height. The scalp
over the bone defect was normal and had an aver-
age amount of hair. The area bulged when the in-
fant cried. Radiographs of the head showed that
this triangular perforation involved the sagittal
borders of both parietal bones. The base of the de-
fect was about 1 cm. above the lambdoid suture
(Fig. 1). The parents were advised that cranio-
plasty might be needed later but that the defect
might partially fill in with time.

She was seen next on March 1, 1960 when the
mother said that the child had developed nor-
mally and that the middle of the hole had filled
in. Towards the end of the year she was to go to
kindergarten and cranioplasty was promised for
about that time.

Examination. The child was normal mentally.
There were two rounded gaps in the skull, one on
each side of the midline in the posterior parietal
region. The scalp was adherent to them but
pulsations were not very marked. The right de-
fect was 3 cm. across and the left was 2.5 cm.
across. There was partial webbing of the inter-
digital space between the 3rd and 4th digits of
both hands and feet.

Radiographs of the skull showed that the de-
fects were of the same size as palpated (Fig. 2).
There was a midline bar of bone between the de-
fects that was crossed by a transverse suture. The
sagittal suture was nearer to the right perfora-
tion but below the transverse suture the sagittal
suture took origin from the inner margin of the
right defect and continued inferiorly to the
lambdoid suture. The lateral edge of the right de-
fect had a small nick in it. The rest of the skull
was normal. Radiographs of the hands and feet
were normal.

Family History. The patient had three siblings
whose heads were radiographed and were normal.
However one brother had had a supernumerary
ulnar finger excised in infancy and radiographs of
the hand were now normal. A sister had a small
exostosis on the shaft of the 3rd metacarpal bone
and some shortening of the ulna on the right. The
heads of the parents were normal and the father's
family had no known abnormalities. The mother
had 13 siblings and of these, two sisters had an
extra ulnar digit and a brother had webbed feet.
A daughter of a normal sister had had severely
webbed fingers for which surgical correction had
been done.

Operation. With the child lying prone the de-
fects were exposed by a curved incision. The
scalp was stuck firmly to the area of the perfora-
tions and several small arteries and veins ran from
the scalp through the defects. These were divided.
A thin flap of fibrous tissue was dissected up from
the membrane covering the defects and this was
continuous with the pericranium. The general ap-
pearance were as indicated by the radiographs
(Figs. 2 and 8). The bone at the edges was thin
and tapered. The gaps were filled in with bone
chips taken from the iliac crest and the peri-
cranial flap was stitched back over the grafts.

Course. Recovery was uneventful and she left
hospital on Nov. 19, 1960. She was seen at follow-
up on Dec. 12, 1960 when the region of the grafts
was firm and painless.

Case 2. E.M.R., a Maori girl aged 5 years, was
admitted to Dunedin Hospital on Jan. 3, 1957.
Shortly after birth it had been noticed that there
was a gap in the bone at the back of the head.
Radiographs, done at Wellington Hospital, con-
formed the anomaly and another small de-
ficiency was found in the skull behind the right

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ear. As far as her guardians knew there had been no change in the size of the gaps over the years. As she was about to start school there was concern about the weakness of the head. She was living away from her parents and nothing was known about the family history.

Examination. The scalp was normal and mobile over the perforations. In the midline of the parieto-occipital area there was a roundish hole in the skull about 5 cm. across. There was another gap about 4 × 2 cm. just above and behind the right ear. These areas pulsed but there was no protrusion of the intracranial contents on straining. The rest of the physical findings were normal and she was of normal intelligence.

Radiographs of the head showed that the defect at the lambda was rounded and the lower margin was made up of the occipital bone. The upper border formed a small peak that ran into the sagittal suture. The edges were irregular and tended to be serrated. Also there was an oval deficiency with rough edges where the right petro-parietal suture joined the lambdoid suture and this spread into part of the occipital bone (Figs. 4 and 5).

The radiographs taken in 1953 were available.

Fig. 1. (left). Case 1. Lateral radiograph of skull shortly after birth. The defect does not go as far inferiorly as the lambdoid suture.

Fig. 2 (right). Case 1. Anteroposterior radiograph of skull at 4½ years. The right perforation is the larger and has a nick in its lateral edge. The central bar of bone has the transverse suture and irregular course of sagittal suture.

Fig. 3. Case 1. Appearance at operation. The peculiarities of the bar of bone are shown with the sagittal suture going into the right defect.
Then the defect at the lambda was more triangular but no smaller in total area than later. The deficiency at asterion had not changed.

**Operation.** On Jan. 7, 1957, with the child lying prone, the perforations were exposed by a curved scalp incision. The scalp was not adherent to the deeper tissues and there were no perforating vessels in either area. A thin flap of fibrous tissue was dissected up from the membrane filling the defects. The defect at the lambda had some serrations to its edges and there was little bevelling of the margins. The superior sagittal sinus could be seen running the length of the gap (Fig. 6). The lateral defect had irregular edges. Both areas were filled in with bone chips taken from the iliac crest and the fibrous tissue flaps were sutured over the grafts.

**Course.** Recovery was uneventful and she left hospital on Jan. 31, 1957. She was seen at follow-up on March 19, 1957. The sites of the bone grafts were firm and painless.

**DISCUSSION**

Small emissary foramina in the parietal bone, situated about 30 mm. anterior to the
lambdoid suture and up to 16 mm. lateral to
the sagittal suture, occur either singly or
bilaterally in some 60 per cent of normal
skulls.\textsuperscript{1,14} The openings are about 0.5 mm. in
diameter and are uncommonly over 1.5 mm.
The foramina contain emissary veins. Rarely
a much larger perforation occurs at about
the same site that may be several centi-
metres across. The early anatomists just
called them perforations.\textsuperscript{5,10} Later they were
entitled foramina parietalia permagna and
after that fenestrae parietales symmetri-
cae.\textsuperscript{7,14,15} This has been anglicised to parietal
fenestrae,\textsuperscript{14} and others call them enlarged
parietal foramina.\textsuperscript{17} The "Catlin mark" is an
eponym sometimes used.\textsuperscript{5,11} The relation of
such defects to parietal emissary foramina is
obscure and they only variably transmit
veins.\textsuperscript{14,16} It would seem better to revert to
perforations and call them congenital perfo-
inations of the skull and cite the place of the
lesion; it is possible that differently placed
perforations may be interrelated.

By far the commonest type of anomaly in
this area is parietal thinness, which is present
in 0.4 per cent of skulls. The consensus of
opinion is that parietal thinness and parietal
defects are not related.\textsuperscript{10,14}

\textit{Description.} In the adult skull perforations
in the parietal bone are parasagittal and
posterior, and while they usually are paired
the gap may be single. More anteriorly
lesions have not been described. The average
diameter is 10 to 20 mm., with an upper
limit of about 50 mm. The largest measured
were: right 57\times43 mm. and left 52\times38
mm.\textsuperscript{11} The holes tend to be oval with the
long axis in the coronal plane. In about 40
per cent of skulls the lesions are unequal in
size. The paired defects are separated by a
sagittal bridge of bone about 10 mm. wide,
along whose length runs the sagittal suture.
This suture may deviate off the midline or
be rudimentary and the inner aspect of the
bone may show a similar divergence of the
superior sagittal sinus. This bony bar may
have a transverse suture.\textsuperscript{2,5,13,15,16} The edges
of the perforations are smooth but there is
sometimes a lateral nick like a rudimentary
suture (Figs. 2 and 3); also the margins are
bevelled with the taper being from the outer
table. The defects are filled with a fibrous
membrane that is continuous with the peri-
cranium and dura mater. It is possible to
split the membrane into two layers. In one
case the arachnoid was pathologically
opaque.\textsuperscript{11} The defects may transmit veins
and arteries.\textsuperscript{11,15} Sometimes separate parietal
emissary veins are present as well.\textsuperscript{14} As in
Case 1 the scalp may be firmly adherent to
the membrane covering the defects.

Several patients have been followed from
birth for several years by serial radio-
graphs.\textsuperscript{5,6,17,16,17} At birth there is a median gap
which may be triangular or round in shape
between the two parietal bones but sepa-
rated from the lambdoid suture by bone
(Fig. 1). Somewhere between 17 and 24
months of age a median bar of bone develops
from the upper and lower margins and splits
the defect into two. This bar is fully formed
by 3 years of age. After this there is no alter-
ation in the shape or size of the perforations.
Measurements taken on Case 1 at birth and
4 years later suggest that the only significant
diminution in the total size of the holes was
because of the appearance of the central
bridge of bone. In relative terms the area of
the gaps gets smaller as the skull grows but
in absolute terms the decrease in size is mini-
mal.

Goldsmith\textsuperscript{6} in 1921 pointed out that there
was a strong hereditary element in some
cases. Since then, several family trees have
been recorded of the condition and up to half
the members of a family may be affected.\textsuperscript{13}
The sex incidence is about equal. No formal
genetic treatment of this problem has yet
appeared but there would seem to be the
possibility of a dominant gene. When War-
kany and Weaver\textsuperscript{18} reviewed the literature
they found an impressive number of cases of
parietal skull perforations associated with
other and often grosser congenital diseases.
Relevant to Case 1 were syndactylism, polydactylism and supernumerary epiph-
yses. However, most of the cases reported in
recent years have been of persons without
other congenital disorders. The incidence in
the general population is less than 1 in
25,000 and the literature contains only about 200 cases. The condition has been found in white and non-white persons.

Case 2 had skull defects at the places occupied in the infantile skull by the posterior (lambda) and posterolateral (asterion) fontanelles. However the size of these gaps was far greater than those of the normal at birth. There was no diminution in size over 4 years. The two perforations had irregular margins and the serrated sutural pattern tended to persist (Figs. 4 and 5). It has always been considered that with traumatic defects of the skull the normal intracranial pulsations exercised a smoothing-off effect on the irregularities of the edge of the bone. In these congenital lesions it seems that the form of the margins is a matter of development and that the intracranial dynamics have little modifying influence. Ingraham and Matson showed a normal pneumoencephalogram with a large defect at the lambda.

There may be some relation between these various defects, as Miller and Keagy illustrated a perforation at the lambda in a family with parasagittal lesions. Kite showed parasagittal perforations where the median bridge had not fully developed but the defects were above the lambdoid suture.

Clinical Findings. Many patients are unaware of the smaller types of parietal defect unless their attention has been drawn to the matter by an incidental radiograph of the skull. Often the parents know of the familial nature of the condition. The larger gaps are apparent in the neonatal period. At first there is marked bulging on crying but within a year or so the areas have become firmer and pulsations are not very obvious. Sometimes the patients complain of headaches and epilepsy has been recorded. Pressure on the defects may be uncomfortable. Radiographs will show the extent of the skull defects and other bones should be examined for anomalies. If pneumoencephalograms are done they are likely to be normal.

Treatment. The natural history of the parasagittal perforations is that they undergo some modification of form in the first 3 years of life and then are stable. Other defects of the skull do not close spontaneously with the growth of the skull. There are no records of any person coming to any harm with such defects, but once parents know that the head is abnormal they exercise such a protective attitude to the child that its mental development may be warped. Under such circumstances cranioplasty becomes mandatory, except for small holes.

The repair of the parasagittal lesions should be delayed until after the child is 3 years of age. As other types of congenital defect do not alter much, in these cases it seems desirable to perform early cranioplasty. The delay in our patients was because of our uncertainties as to the natural history of the conditions.

Tantalum or acrylic cranioplasty has been recommended and the results seem satisfactory. We used autogenous bone grafts but it probably does not matter what method of cranioplasty is used.

Embryology. The parietal bone is developed from two centres in membrane which appear at the 8th week of foetal life. They soon fuse and ossification spreads out in a radial fashion to all margins. At birth there are interosseous fibrous areas at the angles of the parietal bone and the adjacent bones contribute to these areas. These are the fontanelles, the anterior at bregma, the posterior at lambda, the anterolateral at pterion and the posterolateral at asterion. At asterion the fontanelle extends backwards between the supraoccipital and interparietal parts of the occipital bone. While the anterior fontanelle may remain open for up to 2 years the others close at birth. Ossification may be deficient along the lambdoid suture and this is made good by accessory ossifications, constituting sutural (wormian) bones. Each parietal bone develops a transitory cleft on its sagittal border. The two notches make up the parietal (sagittal) fontanelle for emissary veins; this was present in 70 of 95 foetal skulls. At the most this cleft is a small dent or nick at birth. The suggestion that such a feature is commoner in mongols cannot be sustained.

It might be thought that these parasagittal perforations are caused by persistence of
the parietal fontanelle but the defects are much larger than the foetal gaps ever were. In some the parietal emissary foramina co-exist. The subsequent course of the single defect, whereby it becomes bisected by a bridge of bone with a transverse suture, is quite unlike anything that has happened in normal development. All that can be said is that these perforations occur at about the same place as the parietal fontanelle but are a considerable perversion of development. The defects of the kind in Case 2 are in situations of normal fontanelles but again the anomalies are far greater than anything that has happened at any stage in the normal growth of the skull. There is no known cause but there is a strong familial incidence which is suggestive of a dominant genetic effect.

REFERENCES