PRINCIPLES
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DIAGNOSIS
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INTRODUCTION

This book is intended for students of radiology, before and after their diploma examination, and for those clinicians to whom the head is of special concern. There are already a number of text-books in which the skull is mentioned incidentally as part of a description of general skeletal pathology. The principal aim here is to explain more fully the effects of disease of the nervous system, and although generalized non-neurological conditions are included, both for their own sake and as differential diagnoses, much more space and fundamental thought is given to subjects such as raised intracranial pressure than to the local effects of general bone disease.

The author believes that in the process of examining a set of skull radiographs, the system of thought is something of the nature suggested in the central eight chapters—that the eye being directed in turn to different regions of the head, the memory ruminates upon possible causes of any translucencies, sclerotic areas or malformations that may be seen. For this reason the arrangement of chapters is a hybrid of many possible (and on the surface more logical) classifications.

The particular regional subdivision which has been adopted reflects the traditional method of neuroradiology which is to describe in turn the vault, the sella and the base, and then any intracranial calcification which may be present.

There are two main disadvantages of this method. In the first place, a good deal of repetition is unavoidable and will irritate the more knowledgeable (but will perhaps help the beginner). In the second place, it is very difficult to write a text-book of this style in such a way that it is easy to read, for the attention is broken by a multitude of subheadings.

The reader's best approach is to use the subheadings for a preliminary reconnaissance of each main section and not to attempt more until the general scheme has been appreciated.

It is hoped that by cross-references tedious duplication has been avoided; and that the quantity of illustrations will enliven a pictorial memory. There are 640 of them.

The radiology of ears, nose, throat and eyes has much common ground with neuroradiology, and in a fairly simple form these subjects are also dealt with. The radiology of the ear falls into its place in the chapters devoted to the skull base, but the nose, the nasopharynx, the sinuses and the orbits have a chapter on their own. The mandible and teeth have little connection with neuroradiology and are not dealt with.

Three subjects merit individual discussion because of their practical importance. These are Raised Intracranial Pressure, Head Injuries and Radiography (in the last-named chapter a number of normal radiographs may be found, their features are numbered and the key is facing page 350). In order to give the reader an overall view at an early stage, Raised Intracranial Pressure appears first in the book.

The chapter on Radiography will also be found to contain a commentary on the recent report of the study group on terminology published by the International Commission in Neuroradiology. The author hopes that this commentary may be of interest to other neuroradiologists as it attempts to explain why in some points his advice is unorthodox. It also adds practical elaboration where this seems to be lacking in the official text.

The habit of 10 years at one hospital, of making all lateral radiographs centred over the sella, has resulted in very close attention to the condition of the lamina dura—the cortex—of the dorsum and floor of the pituitary fossa; and the reader should be warned that the description of changes in the sella due to raised intracranial pressure goes somewhat beyond the teaching which is current, particularly in asserting that these changes may be recognized and are significant in the old as well as in the young. Present research is directed towards finding out whether similar changes may be observed with vascular hypertension. There is some evidence that this may be so; and in such patients erosion of the lamina dura should be interpreted with caution. A further warning is necessary (it is repeated in the text)—much skull radiography is done without high definition screens, with inexact technique and with film and processing which so interfere with definition that the fine structure of the vault and early erosions or loss of clear-cut shadow of the walls of the sella are obscured. Unless a 'high definition' technique is used

and the film is 'true lateral' to the sella, the appearance of the lamina dura, particularly in the aged patient should be interpreted very cautiously. Indeed, those who are unused to a 'high definition' technique will find it difficult to believe that a diagnosis of raised intracranial pressure can often be suggested on such grounds. It is an interesting reflection that radiology is heading in the direction of histology where there is a place for both high and low power lenses—high and low definition techniques; high definition where radiation dose is of less importance and immobility makes it possible, lower definition where radiation must be cut or speed is essential. With further technical developments these two forms of examination will become more obviously distinct and may require quite different apparatus and demand appropriate processing.

The author apologizes to those who may be inconvenienced by absence of references in the text. They have been omitted in order to make the book less cumbersome for the beginner and not because of a lack of indebtedness to the works of others. The main bibliography undoubtedly has a number of omissions, but ought to be of some value. A short bibliography is also added for the benefit of those with less time for detailed study of the subject. To those whose published works seem to have been incorporated into the pages, but who find no acknowledgment, further apologies are offered.

This is, in a special sense, a personal book and as the illustrations will show, a real attempt has been made to write nothing that has not come within the author's experience. Where this has proved impossible, an indication is usually given.

The reader should be aware of the division between generally accepted fact and statements of the author's original opinion. His ideas about the sella are already sufficiently hedged, and work on the effect of raised intracranial pressure in childhood has been published previously. There does not appear to be any other mention in the literature of the nature of the thickening which takes place in the vault when pressure is relieved. An attempt has been made to simplify and cut through the confusion which surrounds Crouzon's syndrome, cranial dysostosis, hypertelorism and craniostenosis, but there are precedents for this. One perhaps original observation about osteoporosis circumscripta has been added. With these exceptions the book contains nothing which has not already been accepted by at least some other radiologists, and any differences which may be discovered are only those of emphasis or omission.

The book may be read in a variety of ways. The text is short enough to manage from cover to cover in a few hours. Any chapter may be taken separately, but the cross-references should always be followed if such a method is used. For those who need to refer to a few specific diseases, for example meningioma, a special index is given on page 363. Lastly, since the captions to the illustrations are lengthy, a considerable amount may be learned simply by turning over the pages and looking at the pictures.

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ACKNOWLEDGEMENTS

A textbook such as this is written around a collection of cases and could not be begun without the radiographs. They have nearly all been made over a long period of time by a comparatively few radiographers. The care and skill that radiographers exercise is the basis of radiology, and although I do not thank them all by name here, I would like them to know how grateful I am.

My radiological and clinical colleagues at St Bartholomew's Hospital and the National Hospital for Nervous Diseases, Maida Vale, may recognize the pictures I have borrowed. These colleagues, many of them my seniors, are the sources of a great deal of the knowledge I have tried here to organize and I am in their debt. Dr David Sutton, in particular, investigated many of the Maida Vale patients, and Dr R. A. Kemp Harper, Dr George Simon and Dr W. D. Nichol collected a number of the earlier radiographs of the neurological and neurosurgical patients at St Bartholomew's Hospital.

I wish to thank Miss Ileen C. M. Parker who typed the whole original text from dictation, as well as many subsequent drafts, for her long hours of hard and accurate work, and for proof reading, advice and encouragement.

Dr Ian Kelsey Fry has been a great help to me in making constructive criticisms about arrangement and expression in most of the chapters and I have had endless assistance from the staff of Butterworths.

Lastly, I must acknowledge the searches and loans which were made by Dr Henry Bunje, Dr Alan Crispin, Dr John Hodson, Dr Herbert Lehmann and Dr Fritz Starer which resulted in *Figures 47, 49, 100, 102, 107* and *213*.

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CHAPTER 1

RAISED INTRACRANIAL PRESSURE

The intracranial pressure may be raised as a result of a number of different conditions.

(1) A large intracranial mass, such as new growth, abscess or haematoma, raises pressure by displacement and if it appears quickly will have a greater effect than if the brain and blood vessels have time for accommodation.

(2) Obstructive hydrocephalus may follow a partial or complete block of the cerebrospinal fluid pathways by an intracranial mass or congenital anatomical abnormalities or may be post-infective or post-haemorrhagic.

(3) Cerebral oedema may occur around a neoplasm or abscess. It may also be present as a result of encephalitis, cerebral infarction or vascular hypertension.

(4) Craniostenosis (in those severe cases where skull growth falls short of the requirements of the growing brain).

The manifestations of raised intracranial pressure in radiographs of the skull are few in number and not peculiar to particular causes. The changes to be described are acute and chronic with variations in infancy, childhood and adult life. In certain cases of chronic obstructive hydrocephalus, the radiographs show some features which are an aid in the localization of the obstruction.

In the systematic description which follows, it may not be easy for those unfamiliar with neuroradiology to understand which observations are of most frequent clinical importance. They are, in the adult, the changes in the sella turcica, and in the child, the changes in the vault.

About one-third of all patients whose raised intracranial pressure persists for more than 5 or 6 weeks eventually show evidence of its effect upon the routine skull radiographs. Headache and papilloedema are found in many of them but not in all.

INFANCY

Vault

In the ante-natal period and in the first few weeks of life the head is able to enlarge very rapidly, due to spreading of the unformed sutures. The bones of the vault may be thinner than normal, and sometimes if the cause of the rise in intracranial pressure has begun early enough there will be defects in ossification in the parietal or frontal bones—one form of carniolacunia (*Figures 1, 2, 3a* and *78*).

However, only very few faint convolutional impressions are usually visible on the vault bones either in the normal or in infants with hydrocephalus.

Sella

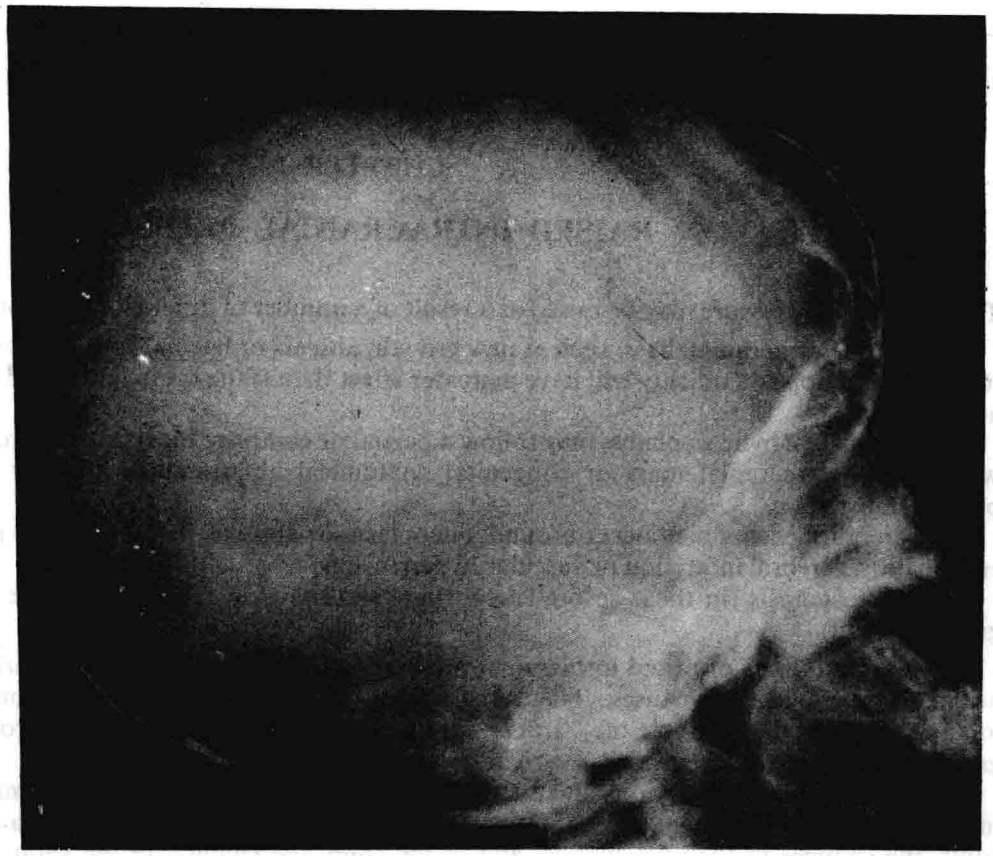
After a few weeks of raised intracranial pressure, a loss of the crisp outline of the lamina dura of the sella turcica may be observed even in infancy but is always less obvious than suture diastasis.

Evidence of the Cause

The commonest cause of raised intracranial pressure in infancy is hydrocephalus, due to a defect of absorption, or to a congenital block at some point in the posterior ventricular pathways. There is frequently an associated meningocoele of the spine.

The skull radiographs may provide some evidence of the position of the obstruction.

In aqueduct stenosis the supratentorial part of the skull may enlarge out of proportion to the posterior fossa which remains shallow. In the author's experience this is most difficult to assess, and in general is a sign of limited value.



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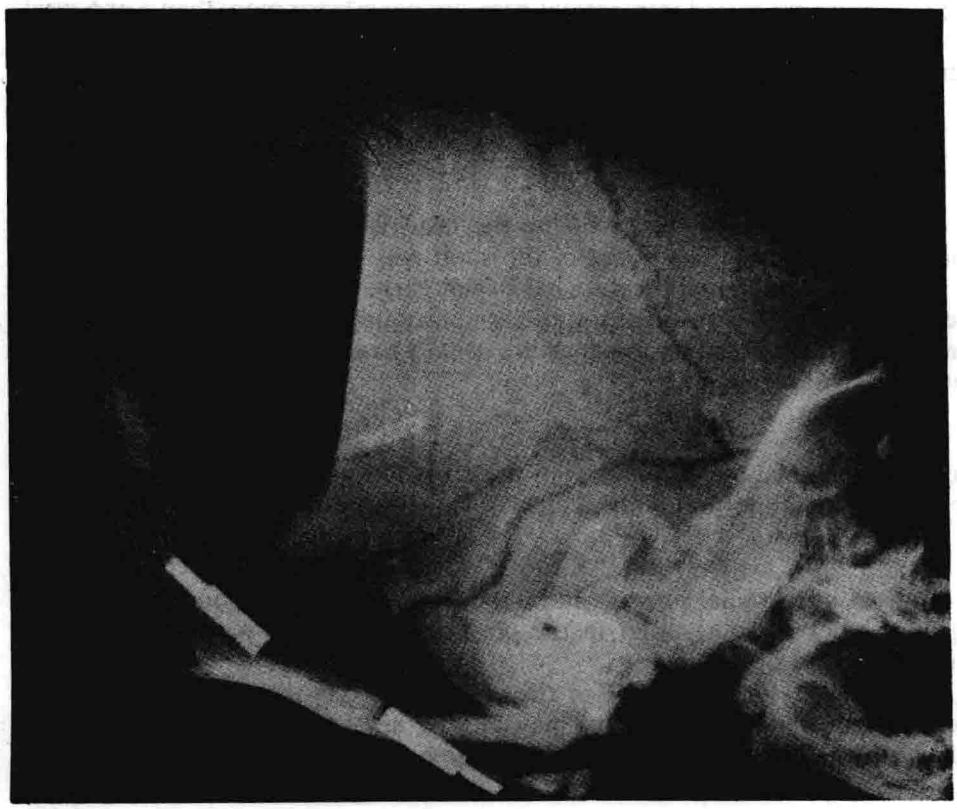

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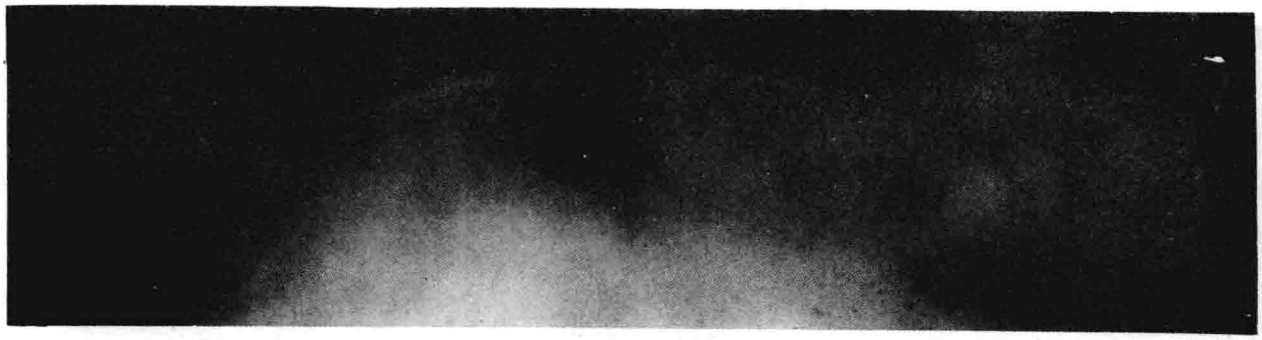
Figure 1. An example of the type of craniolacunia associated with congenital hydrocephalus. (a) Aged 1 week. (b) At 3 months after operation for the meningomyelocoele . and for hydrocephalus. The skull is much more normal though still very thin posteriorly



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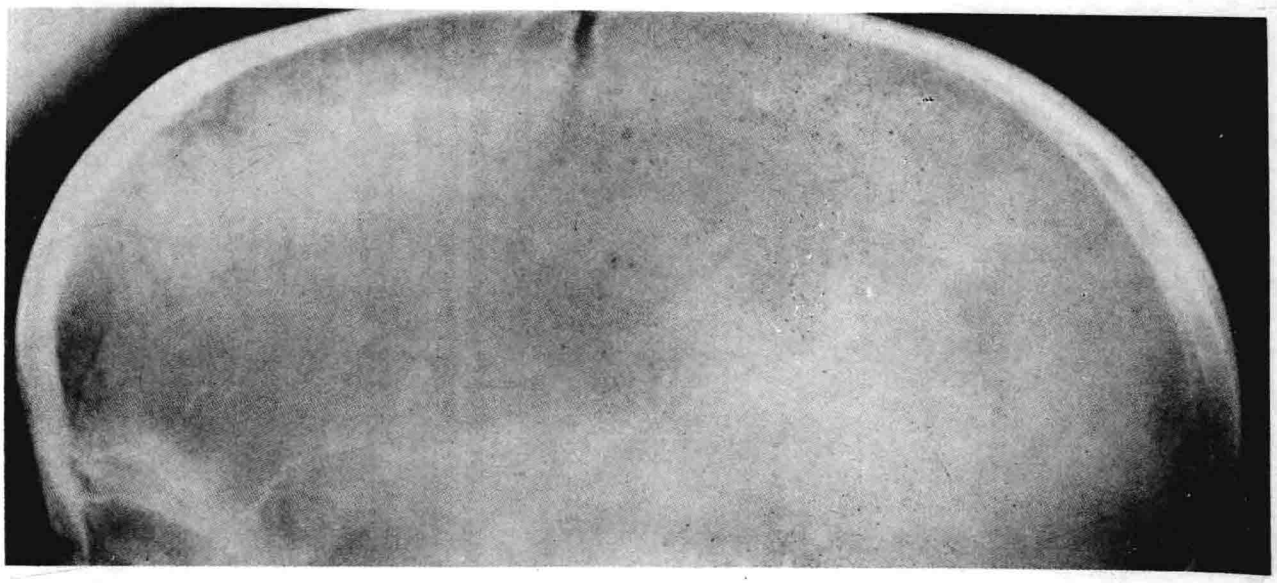

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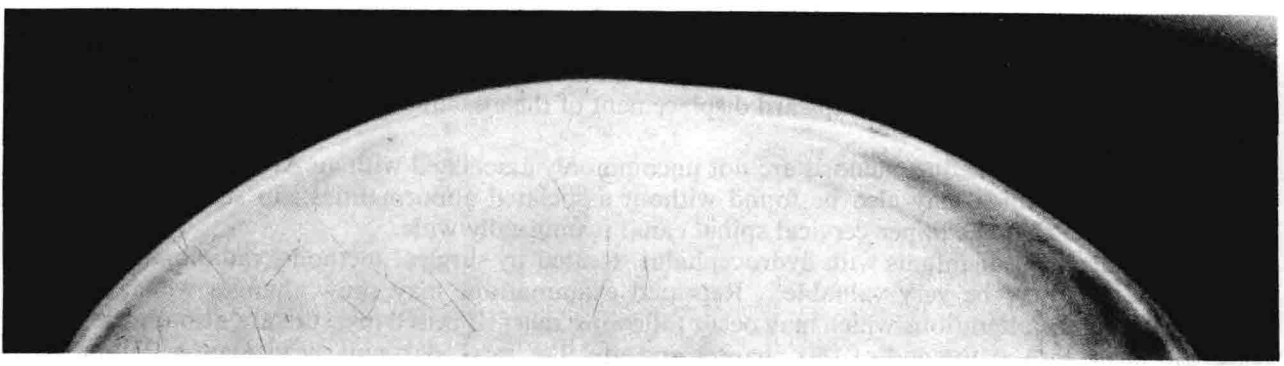


(*b*, ii)

Figure 2. Progress of hydrocephalus. (*a*) *Half axial view. A boy aged 5 months with communicating hydrocephalus. The head is large, the sutures are spread and the bones thin. A theca-peritoneal anastomosis was performed.* (*b*) *The fontanelle became indrawn and the anastomosis appeared to function well for 3 months, but drainage of cerebrospinal fluid into the peritoneum then ceased and the operation was repeated on the opposite side, producing a further improvement for another 9 months. The intracranial pressure then again increased. The lateral and half axial views show how much thicker the bones have become. The sagittal suture has closed but suture diastasis is present elsewhere. There is little attempt at interdigitation, and it may be that this is because closure had almost occurred before this fresh episode of spreading* (*cont.*)

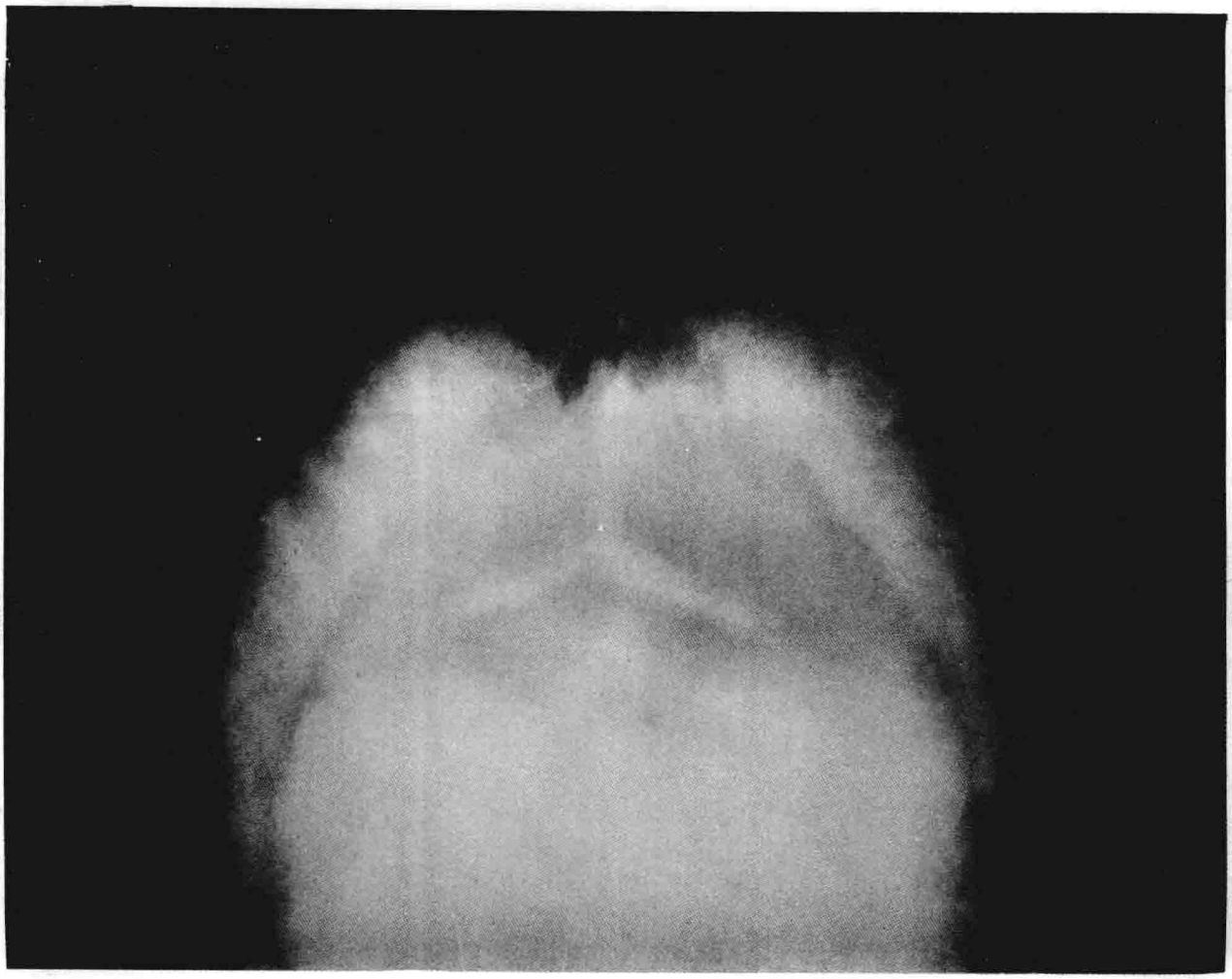


Figure 2 (cont.). (c) One month later there has been a great increase in diastasis; but a further operation reduced the intracranial pressure and the child subsequently improved

In the Dandy–Walker syndrome of atresia of the exits from the fourth ventricle (*Figure 3*), the great dilatation of the fourth ventricle may cause a visible ballooning of the posterior fossa, particularly the squamous occipital bone as well as upward displacement of the attachment of the tentorium (transverse sinus).

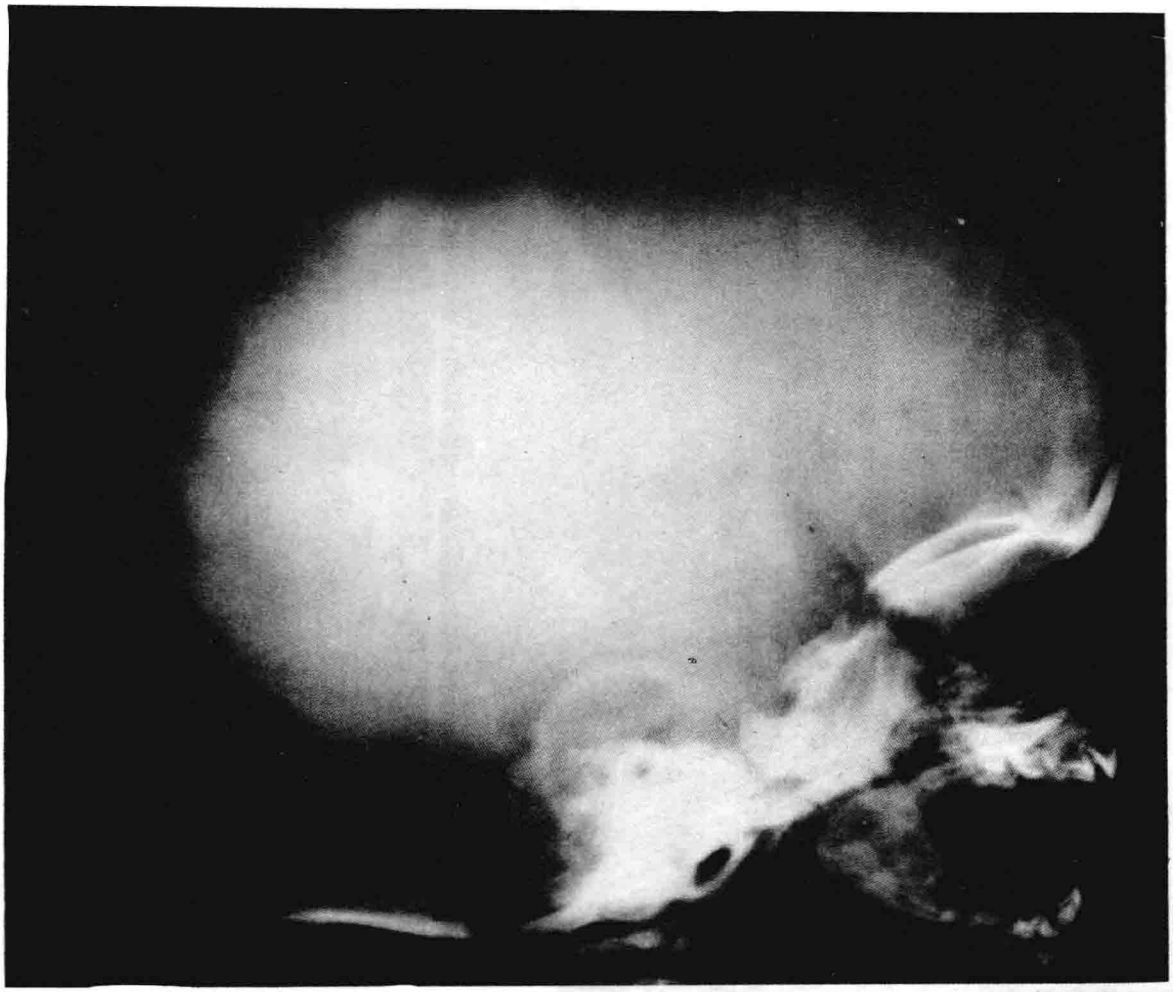
Meningocoele and aqueduct stenosis are not uncommonly associated with an Arnold–Chiari malformation (*Figure 4*) which may also be found without associated abnormalities. In such cases it may be possible to see that the upper cervical spinal canal is unusually wide.

In the management of infants with hydrocephalus, treated by surgical methods, radiological assessment of progress may be very valuable. Repeated examinations may show changes which are bewildering unless the alterations which may occur following relief of raised pressure are also understood. These are dealt with at the end of this chapter and are illustrated particularly in *Figures 25* and *26*.

It is interesting, as yet unexplained but of clinical importance, that neonates with spinal meningocoeles may show increased convolutional impressions (craniolacunia) on their skull radiographs *before* there is any obvious enlargement of the head. In such cases in the author's experience, hydrocephalus soon declares itself.

CHILDHOOD

In childhood, raised intracranial pressure may be of recent onset or long-standing; each has its own radiographic signs.



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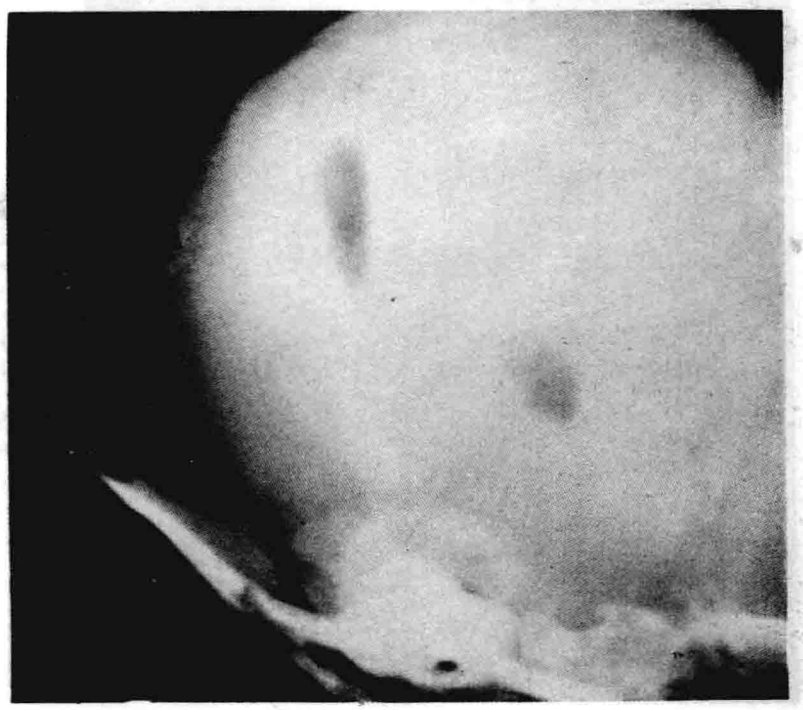
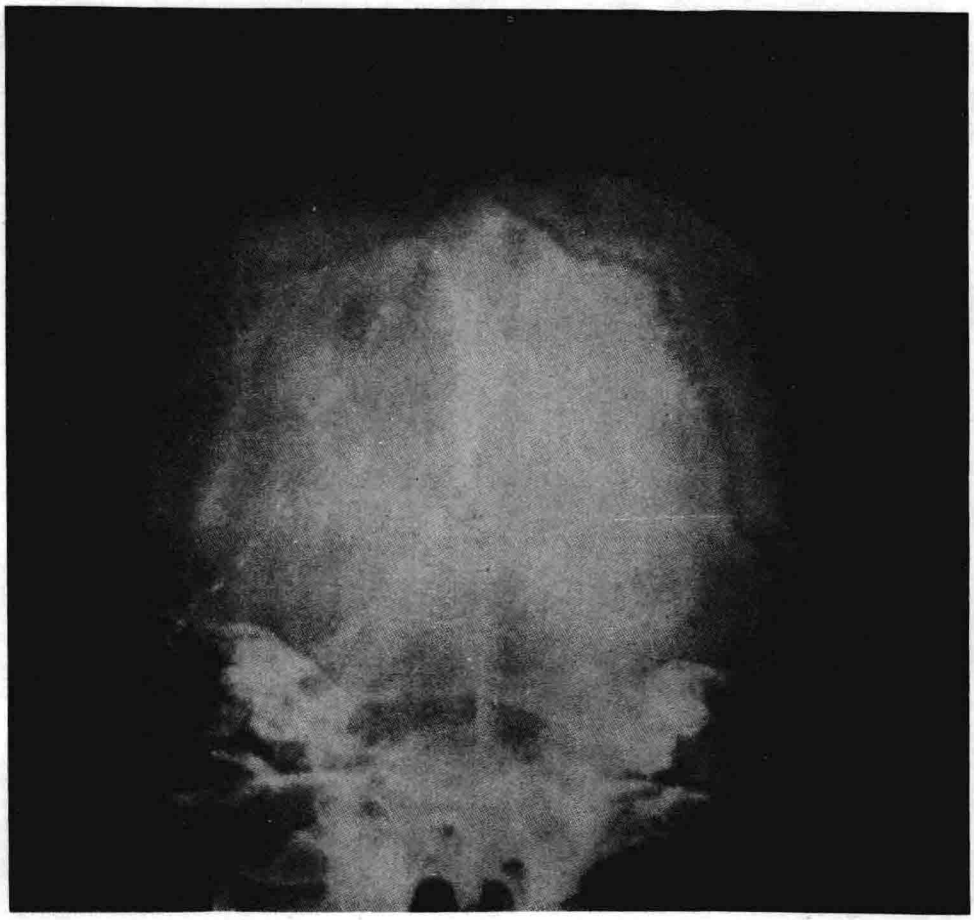


Figure 3. Atresia of the fourth ventricle foramina (Dandy–Walker syndrome). A baby aged 5 weeks who was well for the first 4 weeks of life and then rapidly developed progressive signs of hydrocephalus. (a) A lateral radiograph showing thin vault bones, suture diastasis and bulging fontanelles (particularly the posterior fontanelle). (b) At ventriculography at the age of 8 weeks when bulging of the occiput had become more obvious, air is seen outlining a small part of the enormously expanded fourth ventricle (arrows). This shape of skull in cases of hydrocephalus is characteristic of the Dandy–Walker syndrome (but is not always present). It is accompanied by upward displacement of the tentorial attachment

(*b*)



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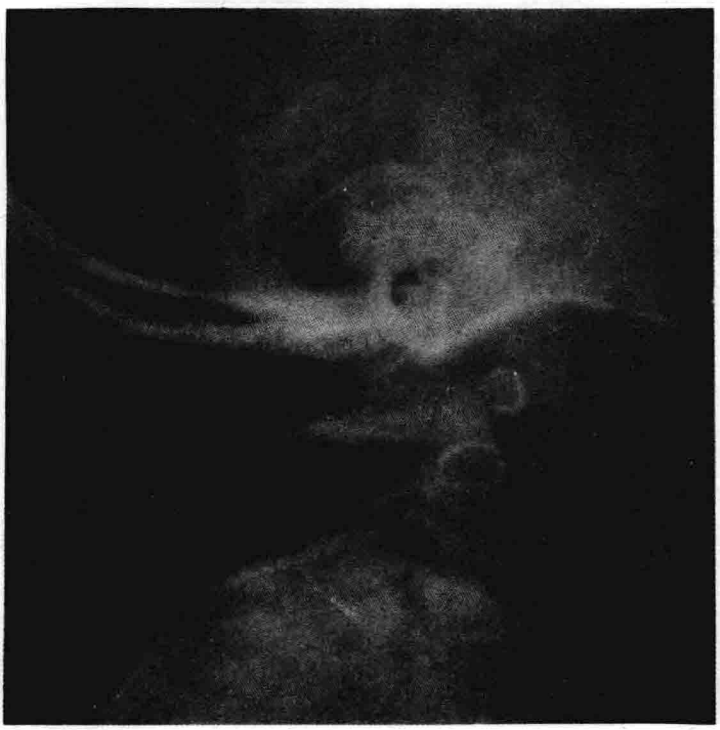


Figure 4a and b. Arnold–Chiari malformation in a child aged 2 years who had had a myelomeningocoele operated on in infancy and later developed hydrocephalus. There is suture diastasis, asymmetrical because the right half of the coronal suture fused during a period of relief from raised intracranial pressure. The foramen magnum is very large and the upper cervical canal is wide. (There is also an unfused posterior arch of atlas.) A large Arnold–Chiari malformation occupied the canal down to C 3

(*b*)

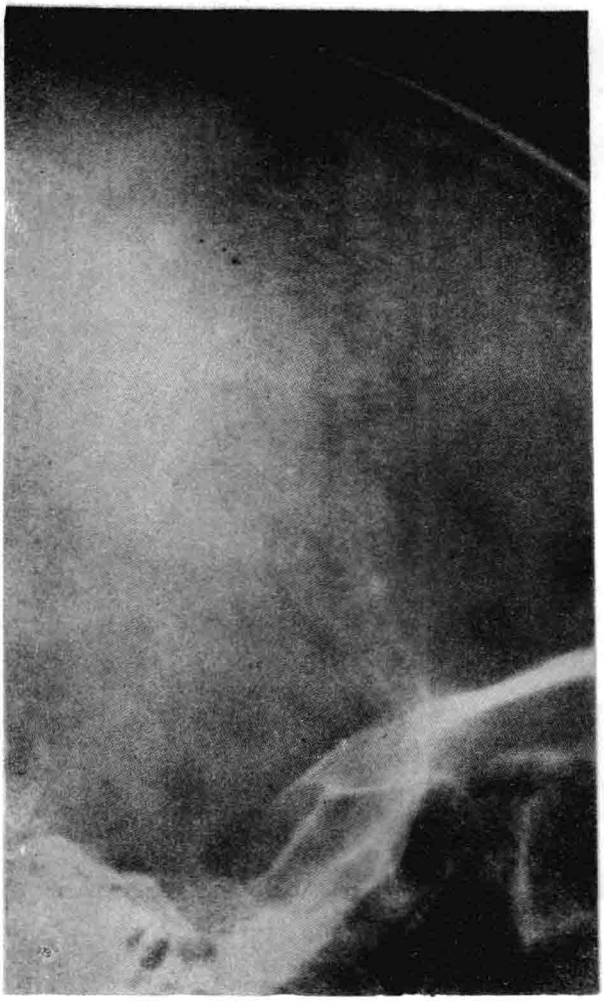


Figure 5. Raised intracranial pressure. A child aged 5 years with a history of raised intracranial pressure of 8 week's duration. The lamina dura of the posterior part of the sella turcica has disappeared (there is also early suture diastasis)

Figure 6. Raised intracranial pressure. Cysti castrocytoma of the cerebellum in a child. The sella shows complete loss of bone detail of its whole floor, in spite of a well-developed sphenoid sinus, and of the front of the dorsum. There is diastasis of stutures, particularly the coronal suture

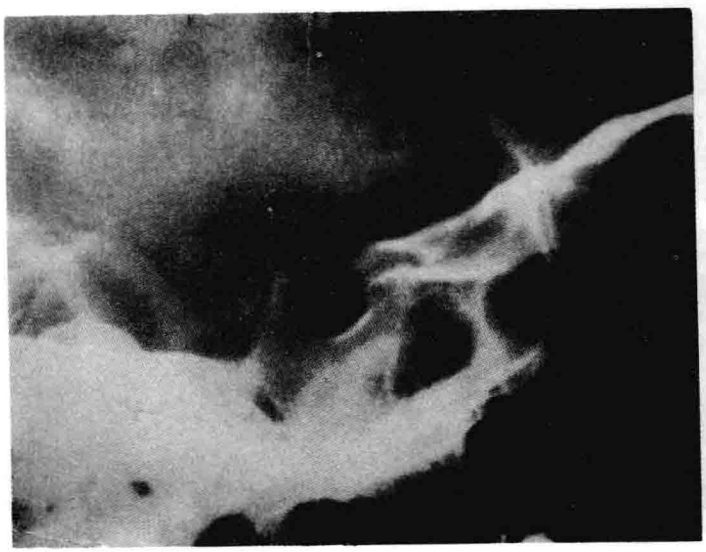
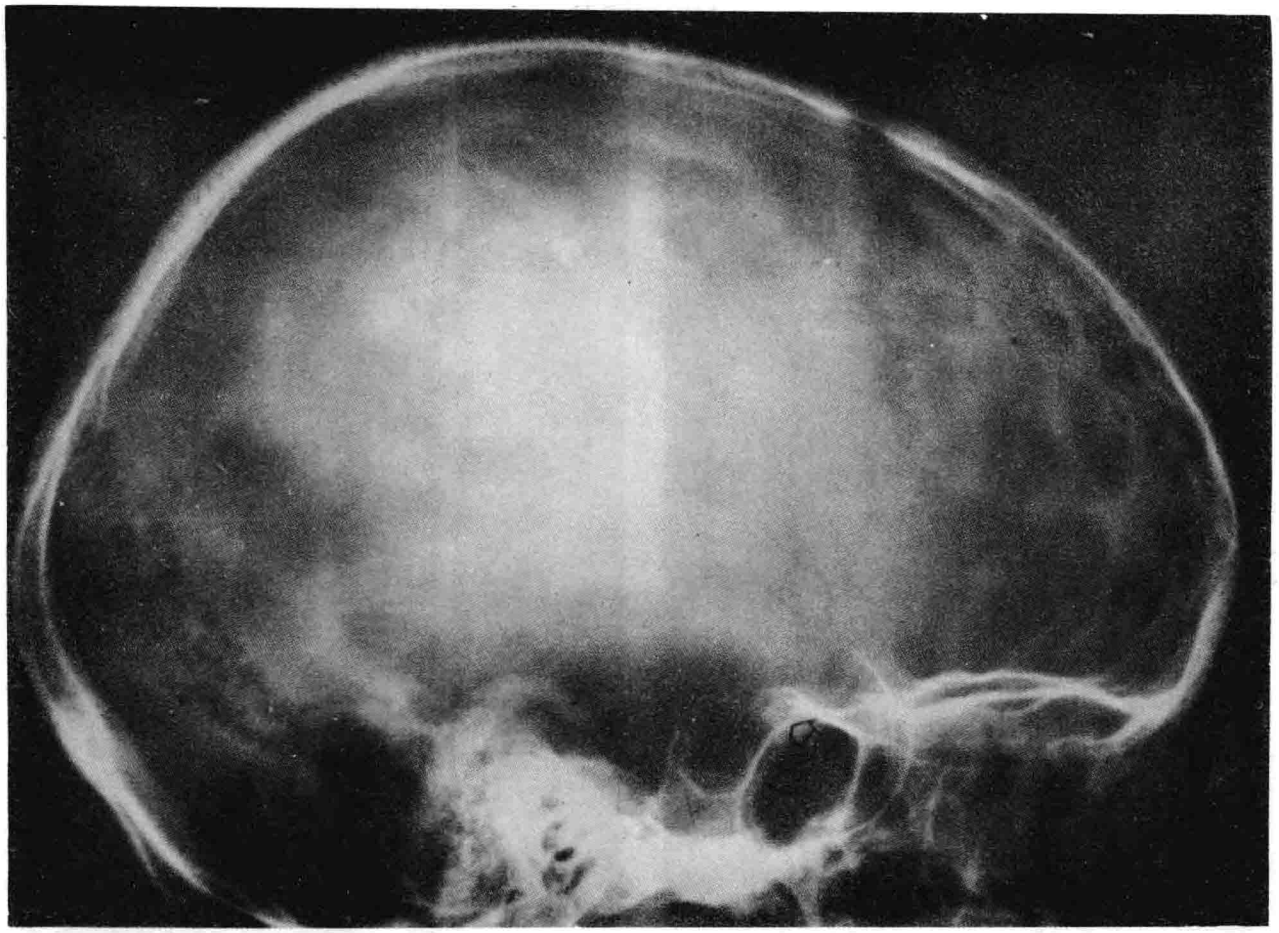


Figure 7. A patient aged 8 years with papilloedema in one half of one retina and cerebellar signs of 5 weeks' duration. The anterior surface of the dorsum sellae has lost its cortex. The tip of the dorsum sellae is more affected than the base



(*a*)

Figure 8. A child suffering from a posterior fossa tumour. (*a*) *Lateral view. The sella appears deep because of a sloping sulcus chiasmaticus. The tip of the dorsum sellae or the posterior clinoids, or both, may be slightly eroded. It is possible that the large and steeply sloping sulcus chiasmaticus may be due to hydrocephalus during active skull growth. The convolutional impressions are very prominent, even over the vertex.* (*b*) *Ventriculogram showing the front end of the ballooned third ventricle* (*closed arrows*) *lying against the sulcus chiasmaticus and the top of the dorsum.* (*c*) *Diastasis of the coronal suture* (*open arrows*)

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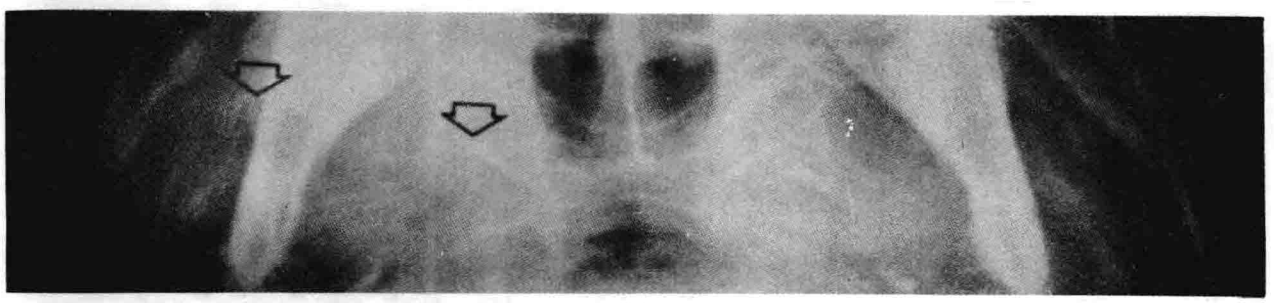

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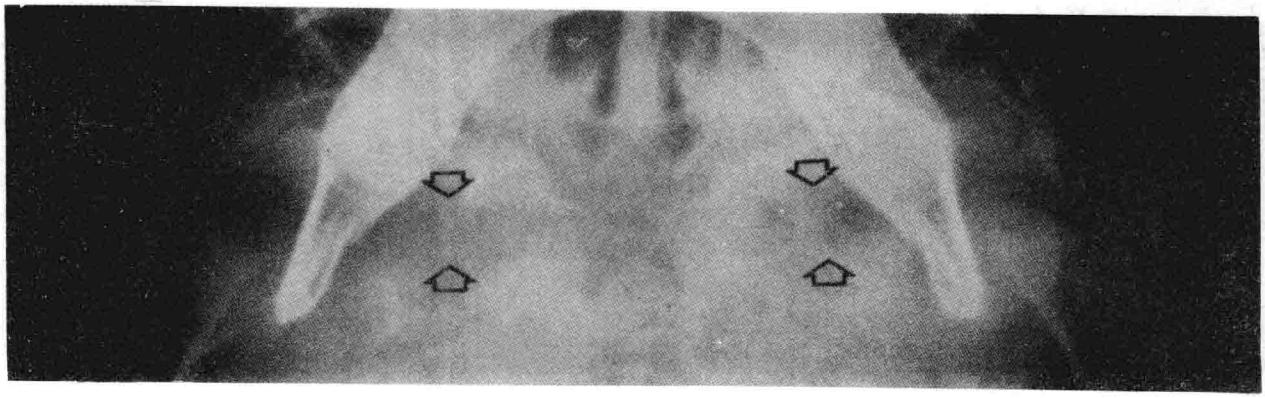
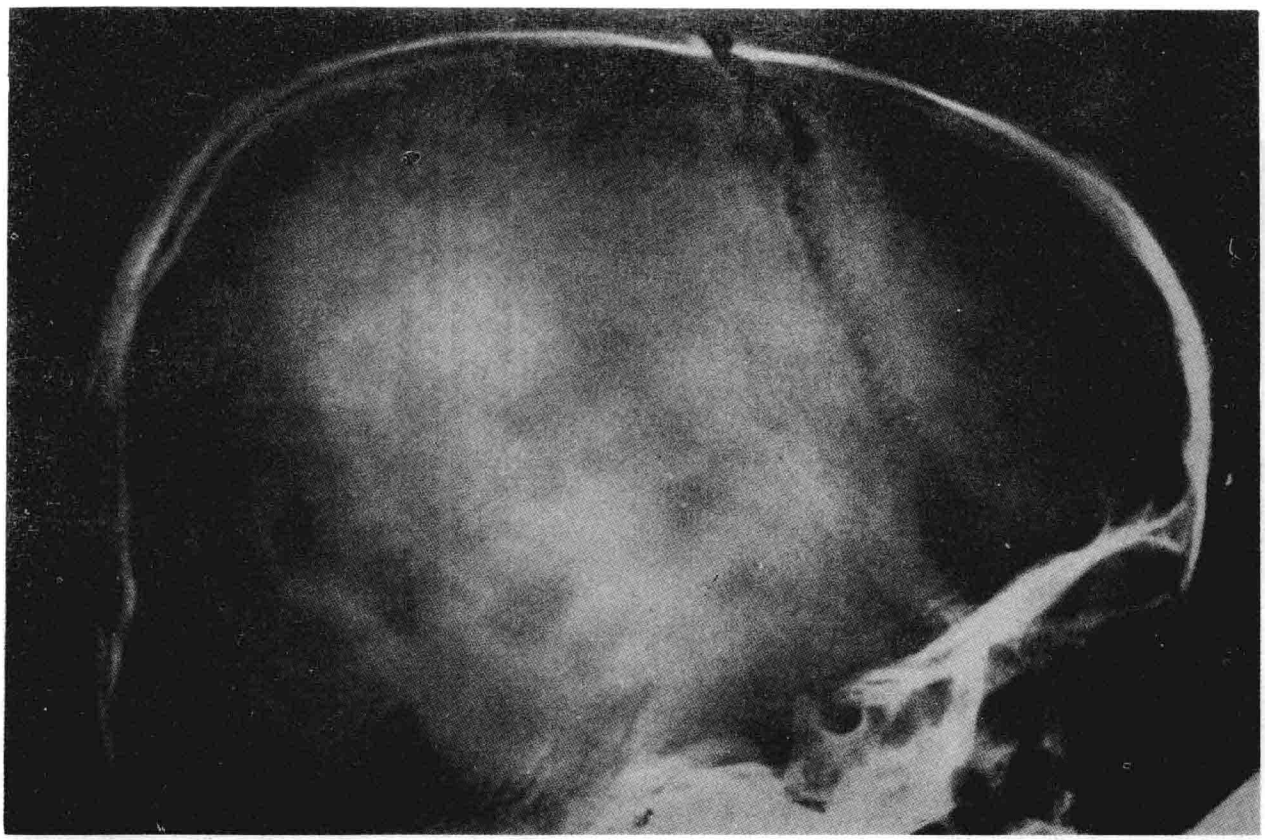


Figure 9. Suture diastasis in a girl aged 1½ years with hydrocephalus controlled by a ventriculo-caval shunt. There is severe diastasis of the coronal suture



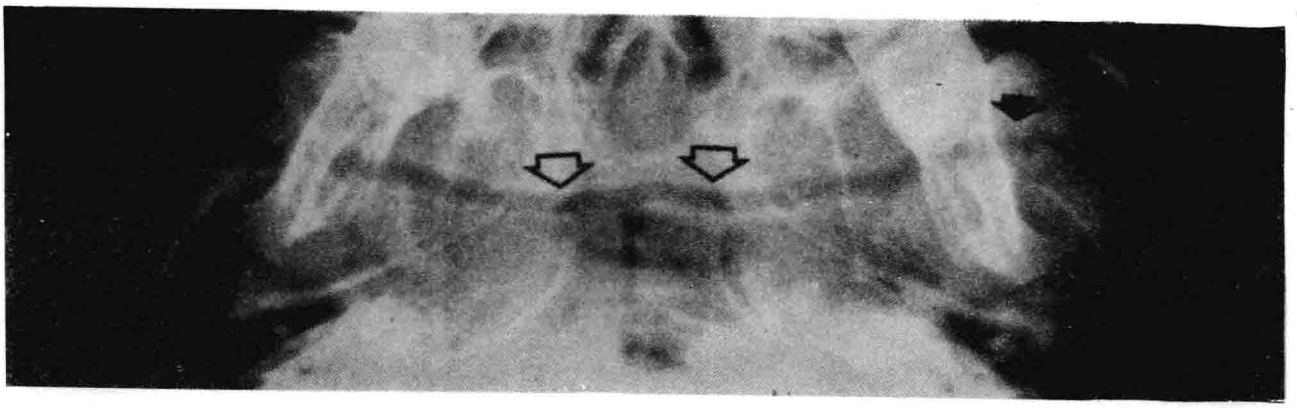
(*a*)

Figure 10. The progress of suture diastasis in a child aged 2 years who had had a theca-peritoneal anastomosis for hydrocephalus soon after birth. He had remained well until 2 weeks before these radiographs were made. He then vomited several times and became listless. (*a*) *The coronal suture in lateral view* (*cont.*)

Raised Intracranial Pressure of Recent Onset (Acute) (*Figures* 5 and 6)

Vault

Up to the age of 8 or 9 years, diastasis of the sutures of the vault occurs easily and may sometimes take place in a few days. After the age of 10 years, suture diastasis becomes uncommon except in very chronic conditions which may have had their origin before the age at which the sutures knit firmly.



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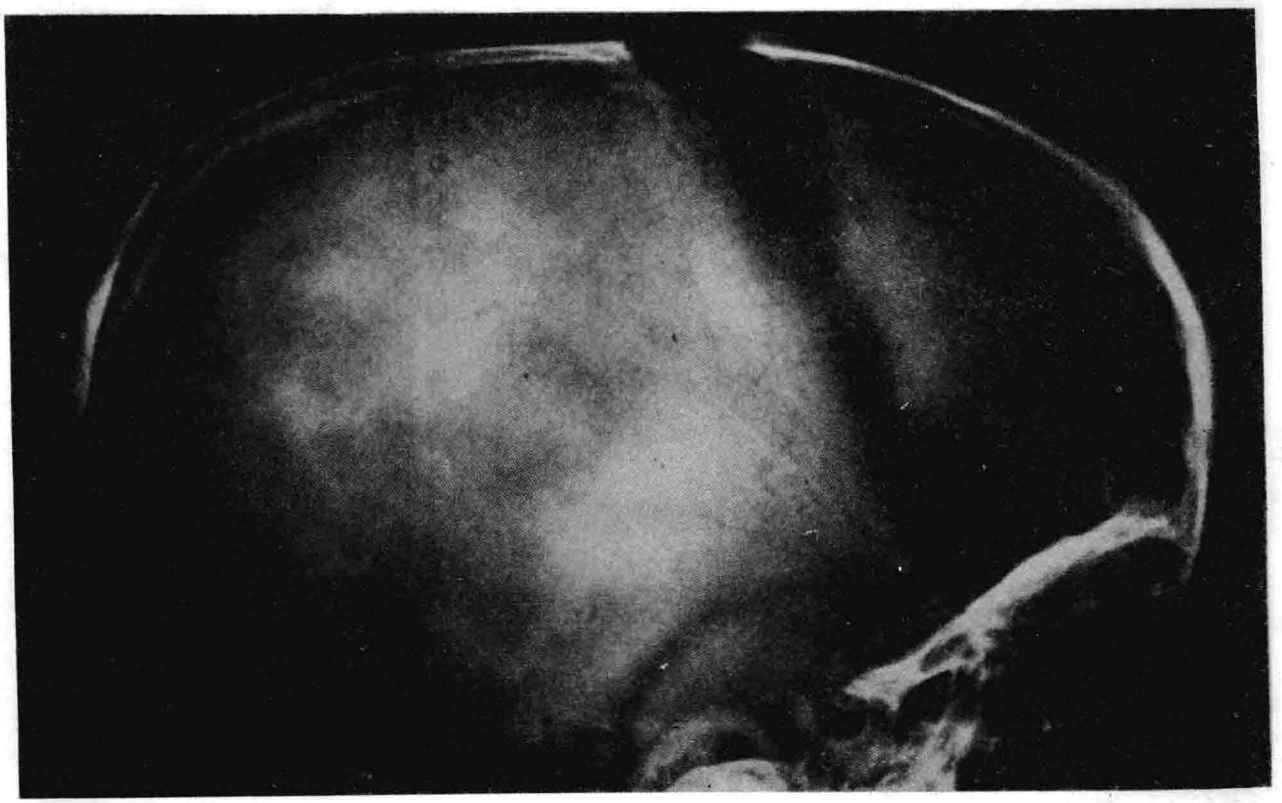

(*c*)

***Figure 10** (cont.). (b) Full axial view. The central part of the coronal suture has never interdigitated (open arrows), but the interdigitations of the more peripheral part are apparent (closed arrow). It was found that the anastomosis tube had fractured, and the shunt was then replaced by one into the epidural space. The patient's head continued to enlarge but his condition improved greatly, and he now (at the age of 9 years) leads an active life. (c) Two months after the previous radiograph the sutures are much wider. Note how 'spikey' the digitations have become as the growing edge of bone attempts to fill the sutural gap*