HYDRAMNIOS AND CONGENITAL FOETAL ANOMALIES

(The Role of Radiology in Diagnosis)

by

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Review of Literature

The association of congenital foetal anomalies and hydramnios is well recognised. Howkins and Lawrie (1939) and Paterson (1944) stressed the importance of radiography in all cases of hydramnios to make a prenatal diagnosis. Macafee (1950) also emphasised the importance of x-ray examination in hydramnios, as a guide to prognosis and future treatment. He analysed 132 cases in whom prenatal skiagrams were available and divided them into two groups. In group I, consisting of 54 patients, x-rays showed gross foetal abnormality either in a single foetus or in one foetus of a multiple pregnancy. Fifty-seven babies were born and out of these 47 had anencephaly and 7 had other gross abnormalities; all these died; there were only 3 normal babies, two of a set of triplets and one of twins survived. In group II, consisting of 78 patients with hydramnios, prenatal skiagrams were reported as normal. There were twenty cases with multiple pregnancies and a total of 100 babies were delivered. Twenty of these were found to have abnormalities like oesophageal atresia, hydro-ureter, congenital heart disease, mongolism and congenital cataract, etc. These anomalies cannot be diagnosed prenatally, but 2 cases of iniencephaly, one each of anencephaly, hydrocephalus and meningocele were missed in prenatal skiagrams in this group. The author feels that with improved radiological technique, most of these anomalies could have been recognised.

Buckingham *et al* (1960) studied 79 cases of hydramnios. Out of 86 infants born, 35, or 42 per cent, were abnormal but only a small fraction of these cases had anomalies which were recognisable on prenatal x-rays. The authors state that in the presence of hydramnios, a normal prenatal foetal x-ray is no positive reassurance, since there remains the chance of gross soft tissue or even bony abnormalities at birth (Buckingham *et al* 1960).

Moya *et al* (1960) found that out of their series of 74 patients with hydramnios, 79 babies were delivered. Twenty-one of these, or 26.6 per cent, had congenital anomalies. Only 11 of these conditions could have been diagnosed prenatally. These consisted of anencephaly in 5, hydrocephaly in 4, microcephaly in one and spina bifida in one. The other 10 anomalies could not have been detected on prenatal skiagrams.

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A larger series of cases with hydramnios studied by Harris *et al* in 1961 showed 47 abnormal babies in 194 cases. Only 28 of these could have been diagnosed prenatally as there were 27 anencephalics and one hydrocephalic foetus. The other 19 foetuses with pyloric stenosis, cerebral agenesis, oesophageal atresia, ventricular septal defect, tracheo-oesophageal atresia and mongolism etc. could not have been seen on prenatal skiagrams.

Present series: From 1st October 1962 to 1st August 1968, a total of 173 patients clinically diagnosed as hydramnios were referred for radiological examination to exclude congenital foetal anomalies. Foetal details could not be made out due to polyhydramnios in 4 patients and these are not included in the study. Out of 169 patients whose x-rays were diagnostic, there was one set of triplets and 42 pairs of twins. Anomalies were visible on prenatal skiagrams in 29 foetuses. (Table I).

TABLE I

Total number of congenitally deformed foetuses—29	
Anencephaly	21
Hydrocephaly	3
Iniencephaly*	2
Holoacardius**	1
Hemiacardius***	1
Cyclops	1

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**Published in the Indian J. of Radiology 17: 18, 1963.

***Published in the Indian Practitioner, 17: 745, 1964.

Anencephaly was the commonest anomaly met with in this series, as in the series reported by other authors. Anencephaly can also occur without hydramnios. The anomaly is easily recognisable on prenatal films and is seen as:

1. Absence of the cranial vault bones; the base of the skull is well developed but irregular.
2. The roof of the orbit is usually deficient; lower facial bones are well developed and two rounded knobs representing the sphenoid bones are seen just above the face.
3. The cervical spine is often defective in development and shortened by lordosis. The malformed base of the skull appearing to be almost at the level of the shoulders.
4. The interpedicular distance is widened; there may be reduction in the number of vertebral bodies and spina bifida of variable extent.
5. The trunk and the limbs are usually normally developed and may show heavy ossification.
6. Due to excess of amniotic fluid, the limbs are displaced away from the trunk instead of the normal attitude of flexion. (Figs. 1, 2).

Soft tissue defects like talipes equinovarus, umbilical hernia, harelip and cleft palate may be associated anomalies (Sarma, 1963) but these are not demonstrable on the prenatal films.

Hydrocephalus was the next common anomaly met with in our series. In this condition, the cranial vault is enlarged with the individual bones separated by gaping sutures and fontanelles. It varies in degree from minor and moderate to major. The prenatal radiological diagnosis is difficult in minor degrees of hydrocephalus, but with good radiological technique it is possible to get at a cor-

rect diagnosis. All the three cases in our series were correctly diagnosed. Although hydramnios is not so commonly found in association with hydrocephalus as it is with anencephalus, it is by no means just a chance occurrence (Prindle *et al*, 1955).

It is important to diagnose the condition prenatally correctly because it may cause dystocia. Radiological features which help in the diagnosis are a variable degree of enlargement of the head, thinning of the cranial bones with face and body relatively small in size; sometimes associated defects like lacunar skull, gross defects of the spine suggestive of spina bifida or meningocele may be visible prenatally as in a case illustrated below. (Figs. 3, 4).

Iniencephaly, a rare foetal anomaly, is incompatible with life and consists of deficiency of the occipital bone in the region of the foramen magnum, spina bifida of considerable extent involving the cervicodorsal spine and extreme lordosis of the spine, with hyperextension of the neck. Usually the diagnosis is made postnatally except in a few cases where it has been made by prenatal x-rays. Both our cases were associated with hydramnios. Prenatal diagnosis is important and the condition has to be differentiated from hyperextension of the head due to muscle spasm in a normal foetus. Since iniencephalus, like other congenital anomalies like anencephaly, is often associated with hydramnios, Howkins and Lawrie (1939) and Paterson (1944) stress the importance of radiography in all cases of hydramnios. A repeat skiagram is usually essential to confirm the persistent hyperextension of the cervical spine. Presence of cervico-dorsal spina bifida, encephalocele and defect in the occiput are diagnostic features.

Usually the labour is uneventful; only 8 out of the 66 cases reported had difficulty in labour. Dorland (1925) reported a case of a large iniencephalic foetus, weighing 9 lbs. 6 oz. causing extreme dystocia necessitating craniotomy. Both the cases encountered by us had typical features of iniencephaly on prenatal x-rays, were correctly diagnosed and delivered normally, though the second case had a large encaphalocele. *Holoacardius acephalus amorphous*, a rare anomaly of the foetus with complete absence of the heart and head, was first described by Beneditti in 1533. Up to 1944, only 63 cases had been reported in the world medical literature (Kappelman, 1944). By the very nature of the anomaly it is found in twins, as the monster depends on the heart of its twin for survival. Usually, the radiological diagnosis is twin pregnancy with one anencephalic monster as in our case. It was only after delivery that a correct diagnosis was made, though on reviewing the films, the upper limbs were also found to be absent. Friedman (1961) reported a similar case where the diagnosis on prenatal skiagerms was twin pregnancy with anencephaly in one foetus. Important features which can help in the prenatal diagnosis are:

i. Usually the upper extremities are also absent.

ii. The malformed foetus is very much smaller than its twin, whereas an anencephalic foetus is similar to

its twin except for absence of the cranial vault.

iii. Anencephaly is much less frequent in twin pregnancy than holoacardius (Friedman, 1960).

Hemiacardiac monster has a rudimentary heart; in our case, two foetuses of unequal size were seen in a woman with acute polyhydramnios and twin pregnancy. The smaller foetus had microcephaly and a small thoracic cage with deficient upper extremities. A correct prenatal diagnosis of acardiac monster was made.

At dissection the monster was found to have a rudimentary heart with single atrium and a single ventricle, malformed thoracic cage and a single deformed upper extremity.

Cyclops is a rare foetal anomaly often associated with hydramnios. Prenatal skiagram in this case showed a very small skull as compared with the size of the body and microcephaly was diagnosed.

Postnatally, the foetus showed fusion of the two orbits into a single diamond shaped cavity lying in the mid line and a large frontal encephalocele. On reviewing the films, the bony defect in the frontal bone was visible, though it was missed earlier.

Comments

In all cases of polyhydramnios, prenatal x-rays are essential, because clinical examination may be inconclusive. In addition to detecting congenital foetal anomalies of the skeleton, the x-rays give information as regards the position, presentation of the foetus, pleural pregnancy and evidence of intrauterine foetal death.

In order to improve the radiological diagnosis, consultation between the obstetrician and the radiologist is essential in each case. Usually both postero-anterior and lateral skiagrams of the abdomen are essential. Use of an abdominal binder helps to compress the abdomen and reduce the girth. Good radiographic technique, with fast screens and films and use of high kilo-voltage technique, reduce the amount of radiation received by the mother and the foetus. Moreover, if meticulous technique is used, repeat films can be avoided.

In some cases, despite all efforts, the quality of radiograph may not be diagnostic due to excess of liquor amnii and it may be necessary to repeat the films after amniocentesis.

In our series of 173 cases, a skiagram of diagnostic quality was obtained in 169; 29 congenital skeletal anomalies were seen in 213 babies, giving an incidence of 14%. In two cases, one of holoacardius and one of cyclops, although abnormal foetuses were reported on prenatal films, the correct diagnosis was only made after delivery.

The incidence of congenital foetal anomalies detected on prenatal films is high enough to justify taking x-rays of all cases of hydramnios, although anomalies other than those of the skeleton cannot be diagnosed on prenatal films of the abdomen unless complicated procedures like amniography are performed.

Summary

One hundred and seventy-three patients with hydramnios were x-rayed. Films of diagnostic quality were obtained in 169. Out of 213 foetuses seen prenatally, congenital skeletal anomalies were correctly diag-

nosed in 29, giving an incidence of 14%. Although soft-tissue anomalies and other organic defects cannot be diagnosed on x-rays, all cases of hydramnios should have prenatal skiagrams done.

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Figs. on Art Paper I