CNS Anomalies – Antenatal Diagnosis in Quantitative Polyhydramnios

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Excess of amniotic fluid is called polyhydramnios. It is suspected clinically and confirmed by sonography. Criteria of an amniotic fluid pocket of 8 or more than 8 cm measured in anteroposterior direction was taken in this study to quantify Polyhydramnios. One hundred patients fulfilling this criterion were included in this study. The degree of polyhydramnios correlated directly that an anomaly would be detected on a prenatal sonogram. Purpose of this study was to evaluate the spectrum and frequency of CNS anomalies in quantitative polyhydramnios. 53 cases were normal where as 47 fetuses were anomalous. A total of 59 anomalies were detected with some fetuses showing multiple anomalies. CNS anomalies were the commonest ones encountered in this study. Neural tube defects, hydrocephalus, encephaloceles, meningoceles, spina bifida, holoprosencephaly, and hydranencephaly were some of the anomalies seen.

Key Words: Polyhydramnios, Fetal Anomalies.

Introduction

The volume of the amniotic fluid varies throughout the pregnancy and is controlled by dynamic interactions among the maternal, fetal and placental compartments. 1 If there is an upset in the balance among these compartments a significant deficiency or excess of amniotic fluid may result known as oligohydramnios and polyhydramnios respectively. Polyhydramnios is suspected clinically and confirmed on sonography when single amniotic fluid pocket is more than 8 cm. Prior to sonographic era approximately 60% of cases of polyhydramnios were believed to be idiopathic, where as 20% were related to maternal illness or diseases such as diabetes mellitus and Rh isoimmunization, 20% were thought to be related to fetal anomalies. 2

Recent observations indicate a more dominant role of anomalous fetal development in the production of polyhydramnios. Neural tube defects 3, 4 including anencephaly, encephalocele, and spina bifida, are among the most common fetal anomalies, occurring in approximately 1.5 : 1000 live births.

The purpose of this study was to address common CNS anomalies in quantitative polyhydramnios.

Aims and Objectives

The aim of this study was to evaluate the spectrum and frequency of CNS anomalies in quantitative polyhydramnios.

Material and Methods

The study was conducted in Services Hospital Lahore in from Feb 2001 to Feb 2002. One hundred patients who met the criteria of having an amniotic fluid pocket depth of 8cm or greater were included in the study. It was necessary that the measured pocket be free of fetal parts and umbilical cord. All observations were made in anteroposterior direction. The cases were taken in the 2nd and 3rd trimester.

Patients were studied with the Toshiba ultrasound scanners equipped with convex transducers of frequency 3.5–5 MHz.

If multiple examinations were performed on a patient the examination with the largest amniotic fluid pocket dimension was included. Measurements were made to the nearest 0.5 cm and then ranked in 2 cm incremental groups from 8.0 cm upward. Obstetrical ultrasound was done and it included fetal number, lie, presentation, gestational age, placental localization and fetal cardiac activity.

Pocket of amniotic fluid was measured in anteroposterior direction and any anomalies seen were recorded.

Results

One hundred patients with an amniotic fluid pocket of 8 cm or greater than 8 cm were included in the study. Of these, 96 were singleton pregnancies and 4 were twin pregnancies. The degree of polyhydramnios correlated directly that an anomaly would be detected on a prenatal sonogram. For the lowest rank group (Pocket depth 8.0 – 9.5 cm), 13 of 51 (24.4%) of fetuses manifested an anomaly, whereas the highest rank (Pocket depth 16.0 cm or greater) carried a risk of 100% (1 of 1) for an anomaly and 2nd highest rank (Pocket depth 14 – 15.5 cm) carried a risk of 88.9% (8 of 9) for an anomaly (Table 1). Of 100 cases studied 47(47%) were anomalous and 53(53%) were normal.

CNS anomalies (Table 2) were most common in our study and a total of 33 CNS anomalies were seen which comprised 55.93% of all the anomalies detected. Anencephaly (Fig. 1) was the commonest CNS anomaly detected which comprised 15 (45.45%) of total CNS anomalies detected.

Hydrocephalus (Fig. 2) was the second most common abnormality found and was detected in 9 (27%) cases.

7 (21%) cases of other neural tube defects such as encephaloceles (Fig. 3) and meningo/myelocoeles and spina bifida were also detected.
One case each of holoprosencephaly and hydranencephaly was detected.

A total of 59 anomalies seen in 45 pregnancies showing some of the fetuses had multiple anomalies.

Discussion
Polyhydramnios is a relatively uncommon complication associated with pregnancy. It occurs in about 1% of pregnancies.5

<table>
<thead>
<tr>
<th>Pocket size</th>
<th>No. of Cases</th>
<th>Normal</th>
<th>% age</th>
<th>Abnormal</th>
<th>% age</th>
</tr>
</thead>
<tbody>
<tr>
<td>8 – 9.5 cm</td>
<td>51</td>
<td>38</td>
<td>74.5</td>
<td>13</td>
<td>25.49</td>
</tr>
<tr>
<td>10 – 11.5 cm</td>
<td>22</td>
<td>9</td>
<td>40.9</td>
<td>13</td>
<td>59.09</td>
</tr>
<tr>
<td>12 – 13.5 cm</td>
<td>17</td>
<td>5</td>
<td>29.4</td>
<td>12</td>
<td>70.6</td>
</tr>
<tr>
<td>14 – 15.5 cm</td>
<td>9</td>
<td>1</td>
<td>11.1</td>
<td>8</td>
<td>88.9</td>
</tr>
<tr>
<td>16 cm and above</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 1: Normal and abnormal fetuses according to amniotic fluid pocket depth (n = 100).

Fig. 1: Anencephalic fetus showing prominent orbits, absence of cerebral tissue more cranially.

Fig. 2: Severe hydrocephalus showing dilated lateral ventricles, thin mantle of cerebral cortex.

Table 2: Various types of CNS anomalies detected.

<table>
<thead>
<tr>
<th>Name of Anomaly</th>
<th>Number of Cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anencephaly</td>
<td>15</td>
<td>45.45</td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>9</td>
<td>27.3</td>
</tr>
<tr>
<td>Encephalocele / Meningo / myelocoele with spina bifida</td>
<td>7</td>
<td>21.2</td>
</tr>
<tr>
<td>Holoprosencephaly</td>
<td>1</td>
<td>0.03</td>
</tr>
<tr>
<td>Hydranencephaly</td>
<td>1</td>
<td>0.03</td>
</tr>
</tbody>
</table>

Table 3: Anomalies seen in 45 singleton pregnancies.

<table>
<thead>
<tr>
<th>Organ System Involved</th>
<th>No. of Cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>CNS</td>
<td>33</td>
<td>55.93</td>
</tr>
<tr>
<td>GIT</td>
<td>7</td>
<td>11.8</td>
</tr>
<tr>
<td>Hydrops fetalis</td>
<td>7</td>
<td>11.8</td>
</tr>
<tr>
<td>Skeletal</td>
<td>4</td>
<td>6.7</td>
</tr>
<tr>
<td>Urinary system</td>
<td>2</td>
<td>3.38</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>6</td>
<td>10.16</td>
</tr>
</tbody>
</table>

The clinical problems associated with polyhydramnios, apart from fetal anomalies, are maternal discomfort, premature labour and many others. Polyhydramnios is suspected clinically and confirmed by sonography.

The results of this study demonstrate that the frequency of anomalies in fetuses increases proportionally to the degree of polyhydramnios complicating the pregnancy. Presence of polyhydramnios was determined according to the amniotic fluid pocket depth greater than or equal to 8cm and then patients were categorized according to the increasing depth of amniotic fluid pocket.

In our study, CNS anomalies were the largest group, which were 33 (55.93%) of 59 total anomalies detected whereas, in review of polyhydramnios by Cardwell6 found CNS defects comprised 50% of congenital malformations.
associated with polyhydramnios. We could not detect rare CNS anomalies because of the smaller sample size.

CNS anomalies were also most common in the study of Baylan, Parisiv et al. Anencephaly was the most common CNS anomaly detected which comprised 15 (45.45%) out of total 33 CNS anomalies detected. Anencephaly was also most common in the study of Desmet et al who concluded that anencephaly represented 48% of CNS anomalies. In our study most of the patients with anencephaly were diagnosed in the third trimester as most patients turned to us in third trimester, whereas, in western studies, incidence of anencephaly is lower in 3rd trimester, because anencephaly is incompatible with fetal life, so once it is detected in the 2nd trimester, pregnancy is terminated in the western countries.

Other open neural tube defects were encephaloceles and meningoceles. Two cases of encephaloceles which were detected in the present study showed a defect in occipital region, one of these was associated with polydactyly and polycystic kidneys suggesting Meckle Gruber Syndrome, which is a lethal autosomal recessive anomaly.9

Two cases of spina bifida (Fig. 5) were detected which were found to be in the lumbosacral area which is the commonest site. One of these was of open type. Open spina bifida was suspected following detection of characteristic associated changes in the head as it is rare for open spina bifida to occur without some cerebral distortion. The characteristic head changes included ventriculomegaly, a lemon-shaped head, and banana sign suggesting Arnold Chiari Type II malformation. BPD and body measurements were also small for gestational age (10).

One case each of holoprosencephaly and hydranencephaly was diagnosed. Holoprosencephaly is a continuum of cerebral malformation resulting from incomplete cleavage of primitive forebrain into two cerebral hemispheres. Our case was of alobar type. There was monoventricle and thalamic fusion. Later on, this was also diagnosed as having chromosomal abnormality in the form of trisomy 13. These chromosomal abnormalities can be found in up to 50% of such cases especially trisomy 13.11

One case of hydranencephaly was diagnosed; there was no detectable cortical mantle which differentiated it from severe hydrocephalus. The thalamus and lower brain stem were intact. No other CNS anomalies were detected in this fetus as hydranencephaly is not associated with other anomalies.12

It is relevant to mention here that sonography remains the primary imaging technique for evaluation of the developing fetus all over the world and especially in developing countries like Pakistan, however, certain limitations exist.

Fig. 3: Encephalocele with visible sulci and calvarial defect.

Fig. 4: Hydranencephaly complete replacement of cerebral tissue with fluid with discontinuous falx and normal thalamus.

Fig. 5: Transverse scan showing V shaped bony defect and longitudinal scan showing a break in the line of echoes from fetal spine: Spina Bifida.
largely due to obscuration of portions of fetal intracranial anatomy caused by reverberation artifacts of the bony calvarium and due to the low sensitivity of prenatal sonography towards malformations of cerebral cortical development.\textsuperscript{14} This lead to the use of an alternative but safe modality like MR and as a result fetal MR as an adjunct to screening sonography has been well documented now\textsuperscript{15} but a debate regarding advantages and disadvantages of fetal imaging with ultrasound and magnetic resonance imaging (MRI) is still going on. Advantages of MR imaging include higher intrinsic sensitivity than sonography to contrast between various cerebral tissues with greater spatial and contrast resolution which can give better anatomic information.\textsuperscript{16} Previous limitations in MR imaging as a result of fetal motion owing to long acquisition times of the conventional spin echo techniques have been overcome by ultra fast MR imaging techniques such as single shot fast spin – echo sequence\textsuperscript{17} and half-Fourier acquired single shot turbo spin-echo.\textsuperscript{18} With these techniques, T\textsubscript{2} – weighted images of the fetus are obtained in less than 1 second per section without image degradation which depict fetal brain anatomy very accurately at various gestational ages and, therefore, considered best sequence. Inspite of all these advances in fetal MR unfortunately, the recommendations in most of the studies are not clear cut. On reviewing the literature we came to a conclusion that both modalities have strengths and weaknesses. In early pregnancy, and where repeated assessment is needed, ultrasound has the obvious advantage. In certain situations where it is difficult to perform Ultrasound, as in the obese patient or a patient with severe oligohydramnios, better images might be obtained by MRI examination. MRI might also identify fetal ischemic lesions early after an insult such as severe maternal trauma or death of a mono-chorionic co – twin.\textsuperscript{19} There is a synergy between ultrasound and MRI for the diagnosis of certain conditions, such as congenital cytomegalovirus infection or cerebellar telangiectasis.\textsuperscript{20} One of the most valuable applications of MR is in the detection of heterotopias and other malformations of fetal cortical development. There is significantly better prognosis in abnormalities such as ventriculomegaly, agenesis of corpus callosum and Dandy-Walker malformations when these anomalies are not associated with cortical malformations.\textsuperscript{21} The sensitivity of sonography regarding such subtle parenchymal abnormalities is low, therefore, MR imaging may be done in such cases. Similarly with the recent advances in the in utero surgery arena, particularly in the repair of myelomeningoceles has necessitated the acquisition of high anatomic resolution of fetal images that can be obtained with MRI. Local conditions and expertise obviously influence the accuracy of both modalities. We, therefore, recommend that keeping in view the recent trends, fetal MRI should be done wherever available for selected patients to rule out any subtle parenchymal anomalies that may be associated with sonographically detectable anomalies and may affect patient’s further management as well as prognosis.

**Conclusion**

CNS anomalies are the commonest anomalies detected in polyhydramniotic pregnancies among which Neural tube defects are the commonest. This study also showed that greater the quantity of polyhydramnios, greater would be the frequency of anomalies. So a radiologist should be even more cautious to look for anomalies when encountered with quantititated severe polyhydramnios.

**References**

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