



## Prenatal Ultrasound Diagnosis of Cystic Hygroma Occurring in Twin Pregnancies

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**Abstract.** Our study reports five cases of prenatal ultrasound diagnosis of nuchal cystic hygroma and early diagnosis for both fetus and mother.

We observed that nuchal cystic hygroma is frequently associated to cytogenetic abnormalities, congenital structural anomalies and non-immune hydrops fetus universalis.

**Key words:** Cystic hygroma, Nonimmune hydrops fetus universalis, Prenatal ultrasound diagnosis

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### INTRODUCTION

The soft tissue of the neck in a fetus is rather difficult to examine in prenatal ultrasonography. This results from the fact that the fetal neck is only a small fragment of its body, and its bent position in the uteral cavity makes the clear picture even more difficult to obtain [Fig. 1]. On the other hand any developmental anomalies present in this region can be seen in USG relatively well because of their fairly big sizes. The developmental malformation which is most often observed and diagnosed ultrasonographically in this region is cystic hygroma [7, 14, 19].

### METHOD

Between 1986 and 1994, 5 cases of cystic hygroma were recognized in a prenatal ultrasonographic examination at the Department of Obstetrics and Gynecology of Kutno Regional Hospital. In all the cases the initial USG diagnosis was confirmed in postpartum examinations of the newborns.

The ultrasonographic diagnosis was performed using a B and K Medical 3535 with a 3.5 MHz convex probe.

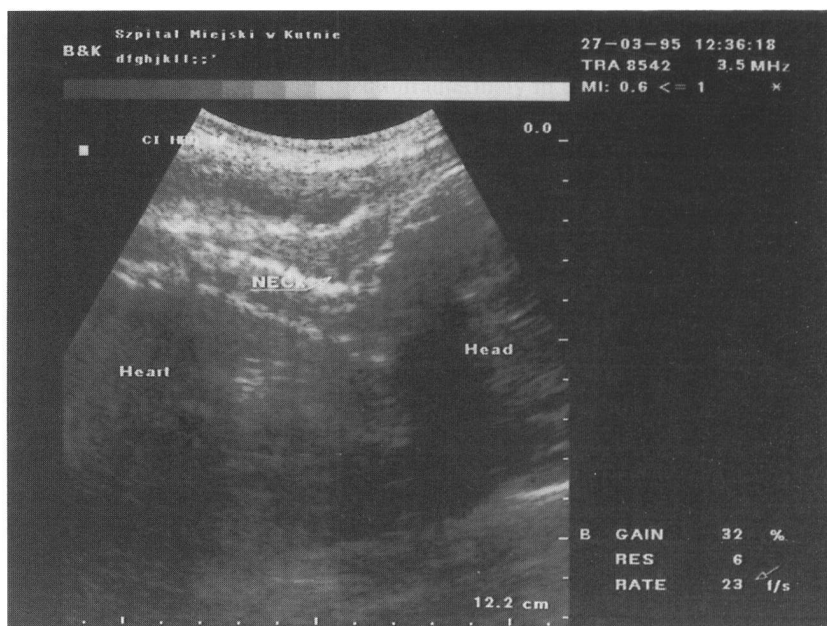


Fig. 1 - Fetal cervical tissue – normal picture.

## DISCUSSION

Cystic hygroma is a congenital abnormality of the lymphatic system, usually observed in the fetal nape [3]. Etiologically the abnormality is believed to result from primary absence or secondary atrophy of connections between the cervical lymph node and the venous system at level of the internal jugular vein, or with the main lymphatic vessel [4, 9, 18]. In consequence the lymph node cannot void, and the collected lymph creates a thin-walled multilocular cystomaous formation in the subcutaneous tissue of the fetal nape [Fig. 2, 3]. The size of this malformation can vary considerably ranging from a small edema of the subcutaneous tissue in the lower part of the fetus's nape to a large cyst spanning from the cranial basis to the lumbar region [11]. The accumulation of serous protein in the cast can be so big that it may result in its deficiency in the circulatory system, and consequently the non-immunological fetal hydrops to develop [11], [Fig. 4]. The collateral circulation between the lymph node and the internal jugular vein, which sometimes builds up can empty the cyst and even make it disappear completely [6, 10].

In the first trimester of pregnancy, cystic hygroma is the most frequently recognized developmental malformation of a fetus observed in screening ultrasonographic examinations [7, 14]. Part of cases are probably undiagnosed because changes, characteristic for a cyst, are not always fully developed at the time. The only early sign, can be the thickening of the tissue in fetus's lower nape with a clear, more than 2 – 3 mm, hypoechogenic area space which separates the skin line from the other tissue of the fetal neck



Fig. 2 - Monolocular fetal cystic hygroma.

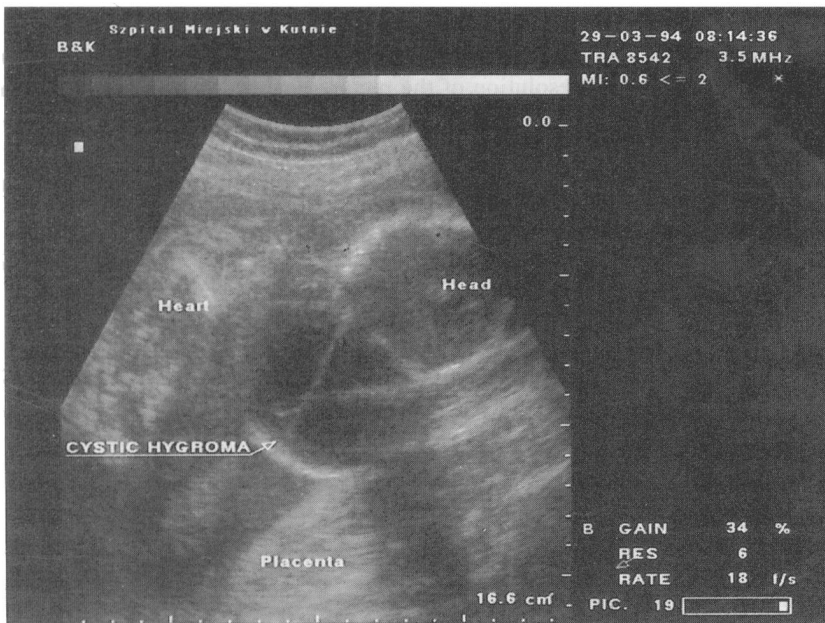
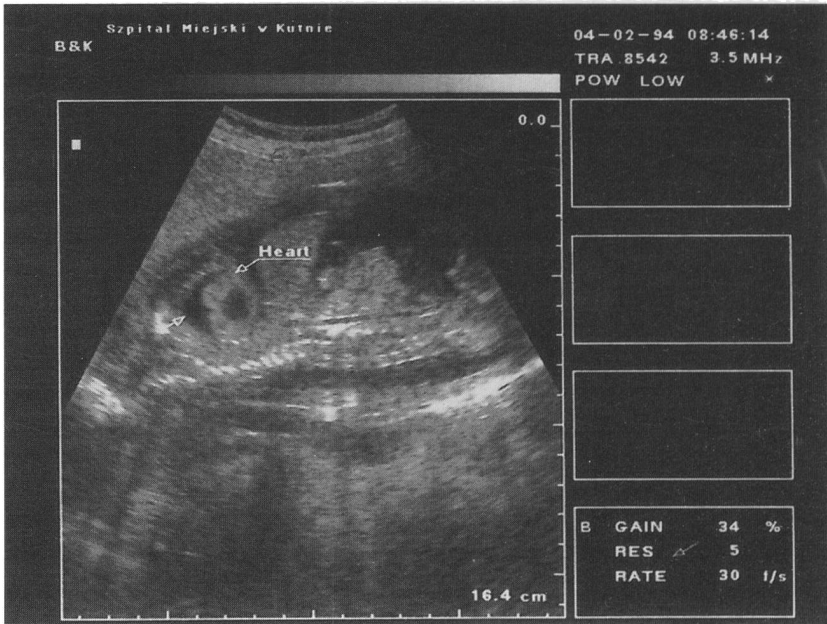


Fig. 3 - Multilocular fetal cystic hygroma.



**Fig. 4 - Fetal hydrops. Free fluid in abdominal cavity and pericardial sac.**

[6, 11, 18]. In case of doubt, it is advisable to re-examine it ultrasonographically after 3 – 4 weeks. Such an examination should be done in order to diagnose decisively and also to assess whether the lesion progresses or regresses. It may also be helpful to use transvaginal probe since the high resolution of the picture enables the tissue to be examined to the smallest detail and, consequently the diagnose to be more precise. The optimal time for USG diagnosis of cystic hygroma seems the end of the first and the beginning of the second trimester of gestation. This is the time when cystic hygroma becomes either a clear hypoechoic space [ $> 6$  mm thick] or a mono/multilocular cystomatous formation in the posterior or posterolateral parts of the fetus's neck [8, 14]. The visualized formation characteristically touches the unchanged osseous system of the fetus's skull and spine [11], [Fig. 2]. Cystic hygroma is often accompanied by hydramnion and fetal hydrops [19].

The differential ultrasonographic diagnosis of cystic hygroma must also include [11, 16, 19].

1. cerebromyelocele;
2. cervical cystic teratoma;
3. branchial cleft cyst;
4. angiomas;
5. mesenchymal sarcoma;
6. thickening of the subcutaneous tissue of the nape often observed in fetal hydrops or 21st chromosome trisomy;

7. second amniotic sac in vanishing twin syndrome;
8. amniotic membrane is not connected with chorion-when the examination is done before the 12th week of gestation.

In the differential examination it is important to recognize the characteristic features of cystic hygroma which are different from those of other craniocervical pathologies [11]. They are:

1. a tumour in the posterior or posterolateral parts of the fetus's neck;
2. a cyst touching the unchanged osseous structure of the fetus's skull and spine;
3. unchanging localization of the formation in relation to the fetus's head;
4. no other solid elements inside the cyst-except for possible partitions.

The differential ultrasonographic diagnosis can be helped by stimulating a fetus to move while observing how the examined structure behaves in relation to the fetus's neck; and by repeating the examination after 3-4 weeks.

The prognosis for fetuses with cervical cystic hygroma is very poor. Most of them – 72.7% – have chromosomal abnormalities, about 60% of them are affected by Turner's syndrome – 45, XO – [1, 2]. In other cases they present trisomies of 13th, 18th, 21st and 22nd chromosomes, and Nonnan's, Cummings' and Rooberts' syndromes [4, 9, 11, 13, 15, 17, 19]. Among other, nonchromosomal malformations which accompany cystic hygroma, congenital heart disease has been diagnosed in 48% of cases and urinary system malformations in 19% [1, 2, 4, 8, 12, 13]. Also in 40%-60% of cases, fetal hydrops developed during the pregnancy [2, 12]. Of 44 pregnancies with cystic hygroma, Cohen et al. [5] had only 3 cases – 7% – of children born alive and only 2 of them had no other malformations and chromosomal defects. The observations presented by Johnson et al. [11] are interesting and of considerable prognostic value. They wrote that when cystic

**Table 1 - USG exponents of cervical cystic hygroma**

1st trimester of gestation	hypoechoogenic space, over 2 mm thick, in the nuchal region of the fetus
2nd trimester of gestation	hypoechoogenic space, over 6 mm thick, or cystomatous formation

**Table 2 - USG exponents of risk degree for fetuses with cervical cystic hygroma**

Lesion/Risk	High	Lower
cyst	multilocular	monolocular
time of appearance	2nd trimester	1st trimester
lesion size	large	small
fetal hydrops	present	absent
malformations and chromosomal defects	present	absent

hygroma developed in the first trimester, the cytogenetic examinations revealed that 39.7% of fetuses were phenotypically normal, and what is more important the lesion regressed spontaneously after 4 – 6 weeks. But when the signs appeared in the second trimester, the percentage of normally developing fetuses was only 25.4%. According to the literature [1, 4, 5, 11], the rate of surviving fetuses with cervical cystic hygroma is from 0 to 7% of all cases. This is the consequence not only of cystic hygroma being concomitant with severe chromosomal defects and malformations but of complications caused by fetal hydrops and prematurity resulting from hydramnion. Among the cases which we observed, 2 were miscarried in the 9th and 14th weeks of gestation, and in 3 the fetuses died intrauterinally between the 30th and 33rd weeks of gestation.

The negative prognosis for fetuses with cystic hygroma particularly depends on the following factors:

1. multilocular cyst;
2. cyst of a considerable size;
3. lesion does not develop until the second trimester;
4. concomitant other malformations and chromosomal defects;
5. non-immunological fetal hydrops.

In each case of a pregnancy with diagnosed cervical cystic hygroma of the fetus a cytogenetic examination should be done and in especially unfavourable situations pregnancy early termination – [2nd trimester] – should be considered.

How much a pregnant woman is endangered depends on how important the lesion of the fetus is. A large cyst may lead to the cephalopelvic disproportion. In our experience there was a case of a fetus that was born dead with a large – 9 cm – multilocular cervical cyst [Fig. 5]. Probably on the way to hospital the mother's uterus body ruptured extensively and the womb had to be surgically removed. Thus the possibility of elective cesarean section has to be considered even when the fetus with cystic hygroma had died intrauterinally before. Such procedure seems justified because of frequent cephalopelvic disproportion and high risk for a parturient if the delivery were through natural passages.

At last we would like to present an example of cystic hygroma which occurs in a twin pregnancy [Fig. 6] from our twin study photocollection.

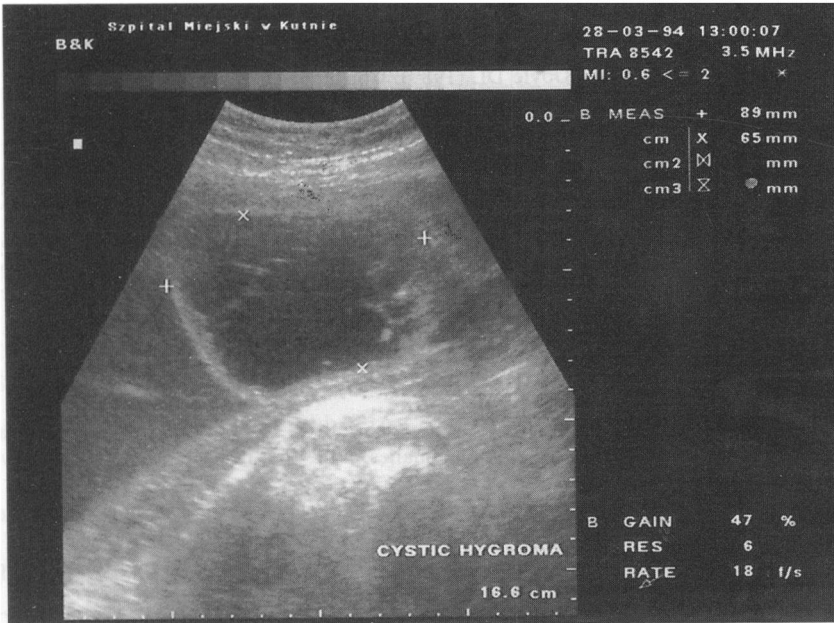


Fig. 5 - Large [9 × 6 cm] multilocular fetal cystic hygroma being an obstacle of labour.



Fig. 6 - Cystic hygroma in twin pregnancy from our twin study photocollection.

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