# Hydrothorax treated in utero and monitored by fetal echocardiography

Wysięk w opłucnej leczony i monitorowany prenatalnie badaniami echokardiograficznymi

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## Abstract

Primary fetal hydrothorax is a relatively rare disorder, occurring in 1 out of 15.000 pregnancies, but connected with high perinatal mortality. In the event of little or no progress of the effusion or stable clinical course, conservative management seems to be the most appropriate course of action. In nearly half of the cases, a placement of only one pleuroamniotic shunt enables a total regression of the primary fetal hydrothorax and, consequently, a favourable outcome. The remaining cases require the placement of yet another shunt. In the case of repeated shunt failure, the prognosis is very poor.

In the following paper we have presented a fetus with a giant re-accumulating hydrothorax after thoracocentesis, treated by pleuroamniotic shunts and monitored by fetal echocardiography and cardiovascular profile score. Despite dramatic fetal presentation and rapid re-accumulation of the hydrothorax, monitoring by fetal echocardiography and cardiovascular profile score proved the repeated fetal needling and double shunting to be safe.

If there are no complications such as premature rupture of the membranes and preterm delivery, fetal echocardiography is a sufficient way of monitoring fetal well-being and the results of intrauterine procedures.

Key words: hydrothorax / fetus / invasive procedures / shunt / echocardiography /

#### Streszczenie

Wysięk w opłucnej u płodu jest rzadką chorobą (1:15.000), ale jest związany z dużą śmiertelnością. W przypadku niewielkiego i nierastającego wysięku postępowanie wyczekujące wydaje się najbardziej właściwe. Założenie shuntu opłucnowego w połowie przypadków doprowadza do regresji wysięku. W przypadkach, w których konieczne jest założenie kolejnego shuntu rokowanie jest bardzo niepomyślne.

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Przedstawiliśmy przypadek płodu z olbrzymim, nawracającym po odbarczeniu wysiękiem w jamie opłucnej, leczonym poprzez założenie shuntu i monitorowanym badaniami echokardiograficznymi i oceną w skali sercowo-naczyniowej Huhty. Pomimo dramatycznego przebiegu i nawrotowego wysięku w jamie opłucnej monitorowanie badaniami echokardiograficznymi i skalą sercowo-naczyniową Huhty wykazało dobry stan płodu poddanego zabiegom prenatalnym.

Jeśli w trakcie zabiegów nie dojdzie do powikłań pod postacią przedwczesnego pęknięcia błon płodowych i porodu przedwczesnego monitorowanie badaniami echokardiograficznymi może odzwierciedlać dobry stan płodu i powodzenie zabiegów wewnątrzmacicznych.

Słowa kluczowe: wysięk w opłucnej / płód / terapia zabiegowa / echokardiografia /

### Introduction

Primary fetal hydrothorax is a relatively rare disorder (1 in 15.000 pregnancies) but it is connected with high perinatal mortality. In the event of little or no progress of the effusion or stable clinical course, conservative management seems to be the most appropriate course of action. Difficulties appear when effusion progresses or rapidly re-accumulates. In nearly half of the cases the placement of only one pleuroamniotic shunt results in total regression of the primary fetal hydrothorax and a favourable outcome. The remaining cases require the placement of yet another shunt. In the case of repeated shunt failure, the prognosis is very poor.

We have presented a fetus with a giant re-accumulating hydrothorax after thoracocentesis, treated by pleuroamniotic shunts and monitored by fetal echocardiography and cardiovascular profile score which have proven the repeated surgical procedure during fetal life to be safe. So far, there has been no data on fetal echocardiography before and after hydrothorax treatment.

#### **Case Report**

Patient GU, 27 years of age, in her second, low-risk pregnancy was referred to our department due to fetus ascites at 23rd week of gestation. We diagnosed a giant bilateral hydrothorax and ascites at 25th week of pregnancy. (Figure. 1A, 1B).

Fetal echocardiography revealed normal heart anatomy with a 9-point value in Cardiovascular Profile Score (CVPS), (1 point was taken away for ascites and hydrothorax). Pleurocenthesis and amniocenthesis for fetal karyotype were performed at 25th week of pregnancy. (Figure 2A).

Fetal karyotype was normal, however, re-accumulation of the pleural effusion in the next 2 weeks took place (Figure 2B).

Therefore, pleuroamniotic shunt was placed at 27th week of pregnancy. Due to ascites and hydrothorax, the CVPS was assessed at the value of 9 based on fetal echocardiography. Two weeks later re-accumulation of pleural effusion occurred, with CVPS of 9. (Figure 3).

At 29th week of pregnancy second pleuroamniotic shunt was placed. Four days after second shunting there was no ascites and only unilateral hydrothorax. (Figure 4A). At 31st week of pregnancy the fetus had again fetal echo and hyperoxygenation test which turned out to be positive. At 34th week of pregnancy there was still only rim of pleural effusion and no ascites (CVPS of 9). At 35th week there was spontaneous rupture of the membranes, however, the newborn was delivered by caesarean section with birth weight of 2200g and Apgar scores of 5 and 7. There was only small pleural effusion in left pleural cavity after the delivery. The newborn was discharged from the hospital 21 days later. After the next two months the weight was 4650g and abdomen, thoracic ultrasound and echocardiography were normal. (Figure 4B).

#### Discussion

Primary fetal hydrothorax (PFHT) may have a highly variable clinical course, from spontaneous regression and survival of the infant without sequelae to fetal or neonatal death. Perinatal mortality in PFHT amounts to 50% [1]. There are a lot of original papers on the topic in question, as well as literature reviews and cases described in Medline data base [2-7]. The conclusions of these studies and research have been very similar for more than ten years now. Despite numerous studies and extensive research, there is only one original paper dealing with fetal echocardiography in PFHT [8]. Bigras et al proved that fetal hydrothorax is accompanied by the compression of the cardiac structures, resulting in altered cardiac hemodynamics and that echocardiographic assessment, including the measurement of the effusion ratio, may be a useful tool in guiding fetal therapy. We have used fetal echocardiography and cardiovascular profile score (CVPS) to assess fetal condition [9]. The fetus had CVPS of 9, despite hydrops. CVPS did not include the diameter of the vessels and ventricles so it did not reflect the cause of hydrops mentioned by Bigras [8].

We took away one point due to ascites. The prognosis is usually poor in fetuses with ascites and functional abnormalities in fetal heart [10]. After first thoracocentesis we observed rapid re-accumulation of the effusion, which was an indication for pleuroamniotic shunt [2]. The CVPS was still 9 points, so we decided to perform pleuroamniotic shunting and two weeks later we observed rapid re-accumulation again. In the event of shunt failure it is acceptable to try a new placement, however, in such cases, the prognosis is very poor indeed. The question how to predict the outcome in such situations remains unanswered. Hydrothorax treated in utero and monitored by fetal echocardiography.



Figure 1. (A) Ascites in fetus at 24th week of pregnancy due to PFHT (CVPS 9), (B) PFHT at 24th week of pregnancy (CVPS 9).



Figure 2. (A) Fetus with PFHT at 25th week of pregnancy, one day after pleurocenthesis (CVPS 9). (B) Rapid re-accumulation of PFHT at 27th week of pregnancy, 2 weeks after pleurocenthesis (CVPS 9).



Figure 3. Another rapid re-accumulation of PFHT at 29th week of pregnancy, 2 weeks after pleuroamniotic shunt (CVPS 9).



**Figure 4. (A)** Unilateral PFHT at 33rd week of pregnancy, 4 weeks after second pleuroamniotic shunt (arrow) (CVPS 9). **(B)** Male baby, 2 months after the birth. Abdomen, thoracic ultrasound and echocardiography – normal.



**Figure 5.** Monitoring and therapy of PFHT (1 thoracocentesis, 2 pleuroamniotic shunt, 9 fetal echocardiography). x – week of pregnancy

y – Cardio Vascular Profile Score

# Conclusions

Despite dramatic fetal presentation and rapid re-accumulation of hydrothorax, monitoring by fetal echocardiography (9 examinations) and cardiovascular profile score proved the repeated fetal needling and double shunting to be safe. (Figure 5).

If there are no complications such as premature rupture of the membranes and preterm delivery, fetal echocardiography is a sufficient way of monitoring fetal well-being and the results of intrauterine procedures.

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