CASE REPORT:

Outcome of infants with hydrocephalus findings on Intra-Uterine Ultrasound (USG) examination at Dr. Soetomo Hospital, Surabaya, Indonesia, in 2015-2017

Lourin Nova1, Mohammad Aldika Akbar1, Agus Sulistyono1, Wihasto Suryaningtyas2, Prastiya Indra Gunawan1
1Department of Obstetrics and Gynecology, 2Department of Neurosurgery, 3Department of Pediatrics, Dr. Soetomo Hospital, Faculty of Medicine, Universitas Airlangga, Surabaya, Indonesia

ABSTRACT

Objectives: to report the outcome of cases with hydrocephalus findings on intra-uterine ultrasound (USG) examinations which happened between January 2015 - December 2017 at Dr. Soetomo Hospital.

Case Report: During the period of January 2015 - December 2017, 20 pregnant women were found who performed an ante natal examination and obtained the fetus with hydrocephalus on intra uterine ultrasound examination. At the time of delivery, out of a total of 20 cases, all babies were born alive, but only 12 babies performed shunting operations with VP Shunt. But at the time of follow up the condition of the baby when the search of this case, found only 7 cases with living conditions. This is probably due to non-routine post-action control to ensure shunt conditions and complications that can result from shunt or other conditions. As seen from the growth and development of 7 surviving children, all cases with normal growth conditions were established based on WHO growth curve and developmental obstacles in all cases with evaluation using DDST II.

Conclusion: Ultrasound examination is useful for determining the fetal prognosis, but for fetoscopic examination and intra uterine operative action remains controversial. Similarly, to determine the exact termination time and delivery mode. Pre-natal counseling and examination is required in mothers with a history of fetal hydrocephalus in previous pregnancies. With routine control is expected better outcomes in fetal hydrocephalus.

Keywords: fetal hydrocephalus; outcome; intra uterine; ultrasound

ABSTRAK

Tujuan: untuk melaporkan luaran kasus dengan temuan hydrocephalus pada pemeriksaan ultrasonografi (USG) intra uterin yang terjadi pada Januari 2015 - Desember 2017 di RSUD Dr. Soetomo.

Laporan Kasus: Selama periode Januari 2015- Desember 2017 didapatkan 20 pasien ibu yang melakukan pemeriksaan ante natal dan didapatkan janin dengan hidrosefalus pada pemeriksaan USG intra uterin. Pada saat melahirkan, dari total 20 kasus yang ada, semua bayi lahir hidup, namun hanya 12 bayi yang dilakukan operasi shunting dengan VP Shunt. Tetapi pada saat follow up kondisi bayi saat dilakukannya penelusuran kasus ini, didapatkan hanya 7 kasus dengan kondisi hidup. Hal ini kemungkinan disebabkan karena tidak melakukan kontrol rutin paska tindakan untuk memastikan kondisi shunt serta komplikasi yang dapat ditimbulkan akibat pemasangan shunt ataupun kondisi lainnya. Adapun bila dilihat dari pertumbuhan dan perkembangan 7 anak yang masih hidup, didapatkan semua kasus dengan kondisi pertumbuhan dalam batas normal berdasarkan kurva pertumbuhan WHO dan adanya hambatan perkembangan pada semua kasus dengan evaluasi menggunakan DDST II.

Simpulan: Pemeriksaan USG bermanfaat untuk menentukan prognosis bayi, namun untuk pemeriksaan fetoskopi dan tindakan operatif intra uterin masih merupakan kontroversi. Demikian pula untuk menentukan waktu terminasi dan cara persalinan yang tepat. Diperlukan konseling dan pemeriksaan pre natal pada ibu dengan riwayat bayi hidrosefalus pada kehamilan sebelumnya. Dengan kontrol rutin diharapkan luaran yang lebih baik pada bayi hidrosefalus.

Kata kunci: bayi hidrosefalus; luaran; ultrasonografi intra uterine

*Correspondence: Mohammad Aldika Akbar, Department of Obstetrics and Gynecology, Faculty of Medicine, Dr. Soetomo Hospital, Surabaya, Indonesia. Phone. Email: dokter_aldi@yahoo.com
INTRODUCTION

Hydrocephalus is a disorder of the central nervous system which manifests as excess cerebrospinal fluid found in the head either inside the ventricular system or the subarachnoid space. The disorder causes the fluid to increase which will further suppress the surrounding brain tissue, especially vital nerve centers. The effect of hydrocephalus is an increase in intracranial pressure which can ultimately be life threatening.\(^1\)

Hydrocephalus is not a single disease but the end result of a broad pathological process both congenitally and as a result of acquired conditions. Therapy should be done as soon as possible. Prognosis of this disease is determined by various factors, including coexisting conditions, duration and severity, as well as patient's response to therapy. Mortality rate of hydrocephalus patients with shunting therapy is still high due to various complications, one of which is postoperative infection. At the moment, congenital hydrocephalus can be diagnosed by intrauterine ultrasound examination and by the time of birth. Diagnosis can be established by clinical signs, supported by sonographic examination, computed tomography (CT) scan or magnetic resonance imaging (MRI). These modalities are intended to examine hydrocephalus in fetus, for possible prenatal diagnosis and further monitoring. Using an accurate ultrasound examination, physician should be able to identify possible therapy interventions that may be taken for the baby after birth, including counseling for the parents related to the baby's prognosis.\(^2\)

Hydrocephalus is an important problem in medicine, as it relates to growth and development since it may disrupt brain growth. Hence, if it is not treated quickly and appropriately, it can cause severe and maybe fatal disruption in growth and development. Statistically, even with good surgical management and medical treatment, only about 40% of hydrocephalus patients have normal intelligence and around 60% of the rest experience significant intelligence and motor function defects. From these statistics, despite getting good neurosurgery and medical treatment, it turns out that about 60% of hydrocephalus patients still experience sequel of significant disorders.\(^3\)

Congenital hydrocephalus occurs in 3 out of 1000 births in the United States and is found even more in developing countries like Brazil, which occurs in 3.16 out of 1000 births. Meanwhile, the incidence of hydrocephalus in Indonesia occurs between 0.2 – 4 out of 1000 births, which is found around 40 – 50% of the entire medical visits or neurosurgery.\(^4,5\)

CASE REPORT

From January 2015 to December 2017, there were 20 pregnant women who received ante natal care at Dr. Soetomo Hospital and were found with fetal hydrocephalus during ultrasound examination in their pregnancy. These pregnant women were referred patients from previous health facilities or obstetrician gynecologist with suspected case of fetal hydrocephalus. Data of the patients were obtained from medical records of Obstetrics Clinic in Dr. Soetomo Hospital from 2015 to 2017, home visit and further follow-up of growth and development in those living children were also carried out.

Table 1. Characteristics of the respondents

| Characteristics                  | Total |
|----------------------------------|-------|
| Maternal age during pregnancy    |       |
| <20 years old                    | 0     |
| 20 – 35 years old               | 15 (75%) |
| >35 years old                    | 5 (25%)  |
| Previous pregnancy history       |       |
| With fetal hydrocephalus         | 0     |
| Without fetal hydrocephalus      | 20 (100%) |
| Primigravida                     | 6 (30%)  |
| Parity                           |       |
| Multigravida                     | 13 (65%) |
| Grande multi                     | 1 (5%)   |
| Midwife                          | 13 (65%) |
| ANC                              |       |
| Obstetrician gynecologist doctor | 7 (35%)  |
| Hypertension                     | 1 (5%)   |
| Diabetes                         | 1 (5%)   |
| Comorbidity during pregnancy     |       |
| Preeclampsia + Diabetes          | 1 (5%)   |
| Obesity                          | 1 (5%)   |
| Others                           | 3 (15%)  |
| No comorbidity                   | 13 (65%) |
In the Table 2, we can see that from a total of 20 patients, only 1 patient was examined for infection factors during pregnancy. The reason behind this finding was because this examination was not routinely performed in the Obstetrics Clinic of Dr. Soetomo Hospital because of its expensive cost. Intra-uterine ultrasound examination of the fetus is one of routine examinations performed on all patients in Obstetrics Clinic in Dr. Soetomo Hospital. In the case of fetal hydrocephalus, in addition to fetal biometry and amniotic fluid examination, special attention to ventriculomegaly width, cerebral mantle thickness and presence of other congenital abnormalities in the fetus were also given.

Table 2. Infection factors examination in respondents

| Examination                              | Total |
|------------------------------------------|-------|
| Toxoplasmosis and CMV examination        | 1 (5%)|
| No examination performed                 | 19 (95%)|
| Total                                    | 20 (100%)|

Table 3. Intrauterine ultrasound examination of the fetus

| Ultrasound examination | Total |
|------------------------|-------|
| Width of ventriculomegaly |       |
| 10-15 mm               | 5 (25%)|
| >15 mm                 | 15 (75%)|
| Thickness of cerebral mantle |       |
| <1 cm                  | 4 (20%)|
| >1 cm                  | 3 (15%)|
| No data                | 13 (65%)|
| Presence of other congenital abnormality |       |
| Present                | 6 (30%)|
| Not present            | 14 (70%)|
| Types of other congenital abnormality found in fetus |       |
| CNS abnormality        | 2 (10%)|
| Heart abnormality      | 0      |
| Digestive tract abnormality | 0      |
| Urinary tract abnormality | 0      |
| Extremities abnormality | 4 (20%)|

Table 4. Pregnancy and infants outcome

| Pregnancy Outcome | Total |
|-------------------|-------|
| Mode of delivery  |       |
| Vaginal Birth     | 2 (10%)|
| Caesarean Section | 18 (90%)|
| Pregnancy age at fetal termination |       |
| <34 weeks         | 2 (10%)|
| 34 – 37 weeks     | 6 (30%)|
| >37 weeks         | 12 (60%)|
| Infants Outcome   |       |
| Sex               |       |
| Male              | 9 (45%)|
| Female            | 11 (55%)|
| Birth Weight (in grams) |       |
| <2000             | 2 (10%)|
| 2000-2500         | 3 (15%)|
| 2500-4000         | 13 (65%)|
| >4000             | 2 (10%)|
| Birth Length (in centimeters) |       |
| <45 cm            | 3 (15%)|
| 45-55 cm          | 17 (85%)|
| >55 cm            | 0      |
| Head Circumference |       |
| <31 cm            | 0      |
| 31-38 cm          | 4 (20%)|
| >38 cm            | 8 (40%)|
| No data           | 8 (40%)|
| Apgar Score       |       |
| 0-3               | 1 (5%)|
| 4-6               | 5 (25%)|
| 7-10              | 9 (45%)|
| No data           | 5 (25%)|
Recommended mode of delivery in fetuses with hydrocephalus is cesarean section. In this case report, there were 2 patients who gave birth normally because of the incident of premature rupture of membrane at 28 weeks' gestation with fetal weight under 2000 grams.

**DISCUSSION**

**Characteristics of hydrocephalus cases in pregnancy**

Risk factors associated with hydrocephalus in the fetus are still uncertain. Previously known several risk factors include infection during pregnancy (toxoplasmosis, CMV, meningitis), family history hydrocephalus (less than 2%), premature pregnancy and low birth weight (which may increase the incidence of intraventricular hemorrhage). The characteristics of pregnant women with hydrocephalus fetuses in Dr. Soetomo Hospital showed that most of them aged between 20 to 35 years, multigravida and none of them had past history of giving birth to children with hydrocephalus or neural tube defect. Landingham (2008) and Hannah (2014) stated in their research that maternal age and parity had no significant consequences to fetal hydrocephalus. From a total of 20 patients, only 1 was tested for toxoplasmosis and CMV to see the infection factor, this is because the examinations cost quite expensive and were not covered by BPJS insurance, hence most mothers refused to do this examination due to financial reasons.

**The diagnosis of fetal hydrocephalus**

Anamnesis done to the pregnant women included questions about her previous pregnancy history,
presence of certain medications consumed, presence of certain diseases in pregnant women and family history of hydrocephalus or neural tube defects.\textsuperscript{9}

Hydrocephalus in the fetus can be detected at the end of the first trimester of pregnancy. However, abnormal dilatation of the fetal brain ventricles is more apparent at 20-24 weeks gestation. In Obstetrics Clinic of Dr. Soetomo Hospital, attending doctors routinely perform ultrasound examination to establish the diagnosis of fetal hydrocephalus. In this case report, we were able to find 2 patients with hydrocephalus fetuses detected on their 24 weeks gestation. However, unfortunately, both patients experienced premature rupture of membranes at 28 weeks gestation which then followed by the infant’s death a few days later. The rest of hydrocephalus cases were detected above 30 weeks of gestation because the pregnant women had just arrived at Dr. Soetomo Hospital after that pregnancy age. This was because the patients were previously under midwife’s care without prior USG examination, while some others had been referred since their second trimester of pregnancy but they just came to the Dr. Soetomo Hospital on their third trimester of pregnancy.

**Pre and post natal prognosis assessment**

**Prenatal prognosis**

The presence of hydrocephalus during the first trimester is a sign of poor prognosis on fetal mortality and its development. Pre-natal prognosis is determined based on intrauterine ultrasound examination of the fetus. Severity of hydrocephalus can be estimated by measuring ventricular width and the thickness of cortical mantle. A finding on cortical mantle less than 1 cm will lead to a poor prognosis. This is due to the fact that the thinner the cortex, the greater the width of the ventricle and as a consequence it can cause further damage to the brain cells and their functions. Hydrocephalus prognosis will become worse if abnormalities other than ventriculomegaly exist. Some disorders that often coexist with hydrocephalus include aqueductus stenosis, spina bifida, Dandy Walker’s syndrome and Chiari malformations. In this study, we found 4 fetuses with cortical mantle thickness <1 cm, 50% were able to survive. This result was slightly worse compared to the study of Peter (1986), who found 78% survival of congenital hydrocephalus patients even though they had cortical mantle thickness under 1 cm without other complications.\textsuperscript{10} Merz (2005) and Paladini (2007) stated that cortical mantle \(\leq 1\) cm lead to a poor prognosis.\textsuperscript{11,12} However, this case report showed that if operative action was carried out immediately after the baby was born, life expectancy would also increase.

**Post natal prognosis**

Ultrasound examination in infants and children is done through an open anterior fontanel. This examination is usually done before the age of 18 months. CT scans can accurately show the size of the ventricles, presence of blood and calcifications, as well as cysts and shunt devices. The advantages of CT scan include clearer picture and it can also determine the prognosis and causes of hydrocephalus. However, many experts are questioning the effects of radiation of CT scans on the brain.\textsuperscript{6,13} MRI can show preoperative information that will determine the success of endoscopic ventriculostomy of the third ventricle and CSF flow postoperatively. MRI examination is done as an alternative examination to CT scan, because the patient is not exposed to radiation and it should provide a whole more information.\textsuperscript{9,14,15}

In our case report, all babies with hydrocephalus who were born at Dr. Soetomo Hospital underwent CT scan except if contraindications were found. Therefore instead of CT scan, MRI or ultrasound examination might be performed. CT scan is a routine examination performed on hydrocephalus infants at Dr. Soetomo Hospital in order to establish the diagnosis and evaluation of hydrocephalus.

**Management of hydrocephalus infants**

Lumbar puncture is a non-surgical therapy for evacuating cerebrospinal fluid (CSF). CSF shunting is a standard measure in the long-term management of hydrocephalus. This procedure puts a ventricular catheter to drain CSF into other cavities in the body, where the CSF fluid can be absorbed. Nielsen (2013) states that hydrocephalus patients who receive surgical treatment has a mortality rate of 10%.\textsuperscript{6} The most frequent complication of VP shunt is infection followed by overdrainage. Recovery period after shunt installation ranges between 3 to 4 days.

In this case series 12 out of 20 hydrocephalus infants were treated with VP shunt. Among 12 infants who underwent the surgery, 6 of them survived (50%), which might be caused by the non-existence of other major congenital abnormalities, immediate VP shunt surgery and adequate postoperative care. The remaining 6 patients died due to postoperative infections. Some infants did not receive operative treatment due to the following causes: 8 infants died immediately after birth due to prematurity, 1 infant had multiple congenital disorders and the remaining 5 infants’ parents refused to do the operative action and decided to request a discharge from hospital. Out of 5 infants whose parents refused operative treatment and requested for a hospital
discharge, 100% were found dead under the age of 3 months. The remaining 1 patient was found with Dandy Walker syndrome and in post-natal evaluation the neurosurgeon advised not to undergo operative treatment, so the infant received only intracranial pressure observation and survived up to the time of this research.

**Expected outcome of infants with hydrocephalus**

In this case report we found that only 40% of hydrocephalus infants survived. This differs with Nielsen's (2013) study which stated that hydrocephalus infants receiving surgical treatment with or without permanent brain damage had a mortality rate of 10%, which means that the survival rate is quite high. Factor that may cause low survival rate of hydrocephalus infants at Dr. Soetomo Hospital includes inadequate postoperative care due to parents' personal request of hospital discharge. Besides, lack of parents' adherence to carry out routine medical check-ups for their babies in order to evaluate the results of postoperative actions highly affected hydrocephalus survival rate.

In this case series, all infants experienced developmental disorder. From a total of 7 children with hydrocephalus who survived, 3 of them experienced gross motor delays, one had gross motor and social personal delays and three suffered from adaptive fine motor, language and gross motor delays which were evaluated using DDST II (Denver Development Screening Test II). These delays are caused by the disease itself which interferes with the development of the brain and also influenced by history of an irregular medical check-up that made observation to infants' development cannot be evaluated better and therefore lacking developmental training.

The expected outcome of hydrocephalus infants is that they can still have life with optimal quality. In this case, growth assessment was carried out using WHO growth chart. Whereas to assess child development, we used DDST II (Denver Development Screening Test II) which was the main revision of the re-standardization of DDST and DDST-R (Denver Development Screening Test Revisited). Observed only from their growth, evaluation of the seven surviving children showed normal growth within weight to height/length limit based on the WHO growth chart.

**Prenatal counseling for future pregnancy**

Up until now there has been no definitive way to prevent the occurrence of fetal hydrocephalus. However, there are several methods to reduce the risk of hydrocephalus in the fetus. First, during pregnancy, prenatal care needs to be done regularly. Second, routine examination and screening of fetal morphology using USG should be carried out at second trimester of pregnancy. Infectious diseases such as toxoplasma and CMV should also be examined if indications are present. In addition, sufficient folic acid intake before and during pregnancy is a must, either through food (spinach, avocado, oranges, etc.) or supplements. Fifth, medical check-ups on cardiovascular-related diseases, such as diabetes, heart disease, increased cholesterol levels, need to be done regularly. Finally, hygiene of cutlery must be maintained to prevent toxoplasma infection which can be related to the occurrence of hydrocephalus.

**Differences in the characteristics of surviving and died infants**

In this case report, out of 20 infants who suffered from hydrocephalus, only 7 survived up to the moment of this research. Among 7 infants who survived, several similarities were found, those similarities were having birth weight more than 2000 grams, being born after 34 weeks of gestation with caesarian section, undergoing immediate operative treatment after birth and receiving good postoperative care before being discharged. This result supports the theory that operative treatment improves the prognosis infants with hydrocephalus. Nielsen (2013) said that hydrocephalus infants who received surgical treatment had a mortality rate of 10%. In this case report, there was only 1 infant who did not receive operative action but nonetheless survived, which was the one with Dandy Walker syndrome. Hatim (2014) stated that therapy for Dandy Walker syndrome was controversial and the mortality rate ranges between 27-50%.

**CONCLUSION**

Based on medical records in Dr. Soetomo Hospital Surabaya during the span of January 2015 to December 2017, 20 cases of pregnancy with fetal hydrocephalus were found. The diagnosis was made by identifying the presence of ventriculomegaly during USG on ante natal examination. Out of 20 cases of pregnancy with hydrocephalus, diagnoses were mostly established at 30 weeks of pregnancy, while in seven cases the babies survived up to the moment of this research. Of the 13 babies who died, prediction of poor prognosis had been speculated since antenatal examination due to the presence of a thin cortex mantle (<1 cm) or presence of comorbidities to ventriculomegaly. At the time of delivery, out of 20 cases, all of the babies were born alive but only 12 of them underwent shunting with VP...
shunt. However, at the time of follow-up, only 7 babies survived and were alive. This was probably due to irregular post-operative medical check-up which supposedly done to ensure the condition of the shunt and complications that might result from shunt installation or other conditions. In regard with the mode of delivery, out of 20 cases found, majority were born through abdomen (18 cases) and 2 cases underwent vaginal birth. According to Chervenak (1986) and Chasen (2001), giving birth through abdomen is the recommended mode of delivery for all cases of hydrocephalus, especially if no other abnormalities are found.\textsuperscript{20,21} Cephalocentesis is only performed on hydrocephalus accompanied by other abnormalities. Whereas, regarding the growth and development of 7 babies who were still alive, all grew within normal limits based on the WHO growth chart with the presence of developmental delays in all cases as evaluated using DDST II. Of the 20 cases found, none of the mothers had become pregnant again. This was because the mothers experienced fear of recurrence of fetal hydrocephalus. Consultation, information and education need to be given before pregnancy planning to reduce the risk of recurrence of hydrocephalus in future pregnancies, including evaluation of toxoplasmosis and CMV infection possibility, more regular antenatal examinations and folic acid supplementation that starts during pregnancy preparation.

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