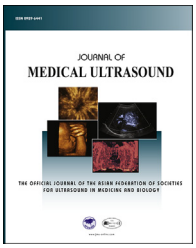
Journal of Medical Ultrasound (2017) **25**, 12—15

Chinese Taipei Society of Ultrasound in Medicine

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.jmu-online.com



REVIEW ARTICLE

Prenatal Ultrasound Evaluation and Outcome of Pregnancy with Fetal Cystic Hygromas and Lymphangiomas

Yen-Ni Chen [1], Chih-Ping Chen [1,2,3,4,5,6*], Chen-Ju Lin [1], Shin-Wen Chen [1]

[1] *Department of Obstetrics and Gynecology,* [2] *Department of Medical Research, Mackay Memorial Hospital, Taipei, Taiwan,* [3] *Institute of Clinical and Community Health Nursing,* [4] *Department of Obstetrics and Gynecology, School of Medicine, National Yang-Ming University, Taipei, Taiwan,* [5] *Department of Biotechnology, Asia University, and* [6] *School of Chinese Medicine, College of Chinese Medicine, China Medical University, Taichung, Taiwan*

Received 17 January 2017; accepted 21 February 2017
Available online 17 April 2017

KEYWORDS
cystic hygroma,
hydrops fetalis,
lymphangioma,
nuchal translucency

Abstract Cystic hygroma is a type of lymphangioma, which is a vascular anomaly associated with lymphatic malformations and formed by fluid accumulation mainly located at the cervicofacial and axillary regions. Cystic hygroma is mostly located in the neck (75%), followed by axilla (20%), retroperitoneum and intra-abdominal organs (2%), limbs and bones (2%), and mediastinum (1%). It is often associated with chromosome aneuploidies, hydrops fetalis, and even intrauterine fetal demise. The prognostic factors of the fetal cystic hygroma or lymphangioma are chromosome abnormalities, hydrops fetalis, septations, or thickness of the cystic hygroma and are associated with other major malformations. Prenatal managements including ultrasound serial follow-up, magnetic resonance imaging, or even intrauterine injection of sclerosing agents are suggested. For fetus with the risk of airway obstruction at delivery, ex utero intrapartum treatment is also indicated. Detailed prenatal counseling is necessary for better neonatal outcome.

Conflicts of interest: The authors have no conflicts of interest relevant to this article.

* Correspondence to: Dr Chih-Ping Chen, Department of Obstetrics and Gynecology, Mackay Memorial Hospital, 92, Section 2, Chung-Shan North Road, Taipei, Taiwan.
E-mail address: cpc_mmh@yahoo.com (C.-P. Chen).

http://dx.doi.org/10.1016/j.jmu.2017.02.001

Introduction

There are different nomenclatures of cystic hygroma. Cystic hygroma is a type of lymphangioma, which is also called nuchal edema or increased nuchal translucency (NT) during the first trimester. It is a vascular anomaly associated with lymphatic malformations and formed by fluid accumulation mainly located at the cervicofacial and axillary regions [1]. It is mainly located in the neck (75%), followed by axilla (20%), retroperitoneum and intra-abdominal organs (2%), limbs and bones (2%), and mediastinum (1%) [2,3]. The thickness is usually ≧ 3 mm. The incidence of nuchal cystic hygroma is about 1/6000 at birth and about 1/750 in spontaneous abortion [4].

It is not only associated with lymphatic malformation but also with chromosome aneuploidies, hydrops fetalis, and even intrauterine fetal demise (IUFD). The prognosis is often considered poor. However, cystic hygroma could be transient in ultrasound findings. It might regress during pregnancy because of recanalization or the formation of collaterals [5].

Here, we will review the diagnosis, factors that affect the prognosis, and the possible management of fetal cystic hygroma for better perinatal outcome.

Prenatal ultrasound findings

Ultrasound findings of cystic hygroma include thin-walled and serpiginous or multiseptated intradermal fluid collections which are often found at cervical regions [6]. Ville et al [7] defined nuchal cystic hygroma as an area of sonolucency in the soft tissue of the occipital region, consisted of two symmetrical cavities completed separated by a midline septum, with or without the internal trabeculae (multiloculated cysts). NT is the presence of unilocular collection of nuchal fluid ≧ 3mm^3. The thickness of the cystic hygroma is measured at its widest part from the intact skull or skin at the transverse view. Prenatal ultrasound of cystic hygroma may show increased nuchal thickness (≧ 3mm), with or without septation at the neck region or thin-walled, sonolucent, and multilocular structure at other regions (Figures 1 and 2). Color Doppler may show no obvious internal flow which can be distinguished from

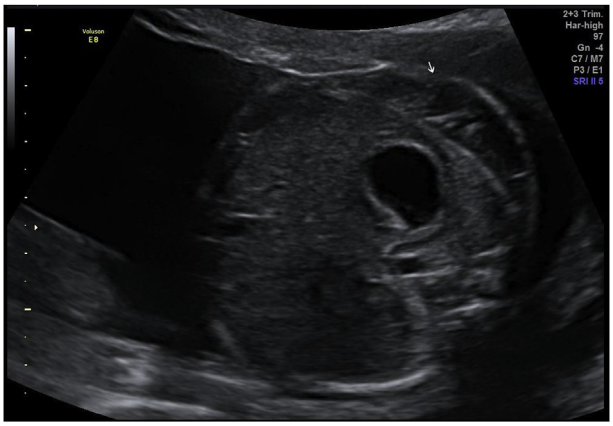


Figure 2 Lymphangioma over the abdominal area (arrow).

hemangioma (Figure 3). Besides, the color Doppler ultrasound is also effective for the detection of intralesional hemorrhage. It may show pulsations from the septums toward the cysts [8]. Furthermore, differential diagnosis should also include encephalocle or cervical teratoma. Head and spine morphologies should be further evaluated for suspected neural tubal defects. Polyhydramnios is also an indicator of neural tube defect but seldom seen in cystic hygroma.

Cystic hygroma in the anterior triangle of the neck is often associated with airway compression, which needs more aggressive intrapartum management. Further image evaluation using MRI is also recommended prior to birth for definite tumor size and infiltration pattern.

Common associated ultrasound findings are hydrops fetalis, cardiac malformations, and skeletal abnormalities. Cardiac malformations are the main anomalies detected in fetuses with normal karyotype (62.2—72.7%) [9]. Other major malformations, such as hydrocephalus, arthrogryposis, agenesis of corpus callosum, pes equinovarus, diaphragmatic hernia, amniotic band syndrome, mesomelia, and bilateral hydronephrosis, were also reported in patients with septated cystic hygroma and normal karyotype [10].

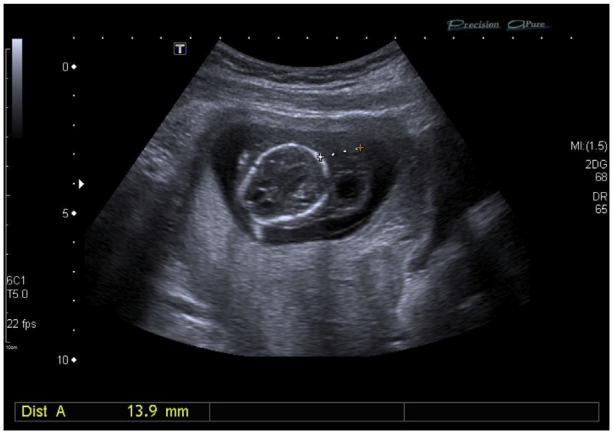


Figure 1 Neck cystic hygroma with a thickness of 13.9 mm.

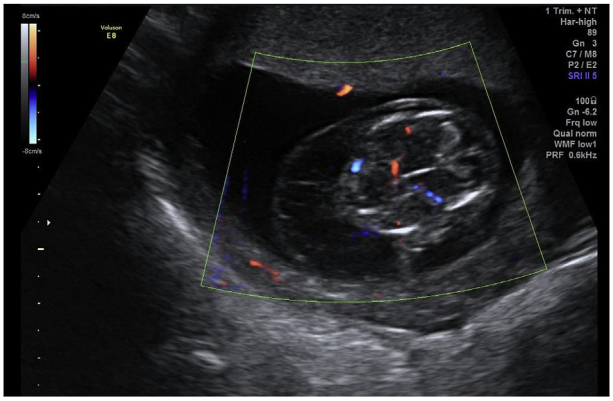


Figure 3 Color Doppler ultrasounds of cystic hygroma showed no internal flow.