

# Prenatal diagnosis and follow-up of 14 cases of unilateral ventriculomegaly

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## ABSTRACT

**Objective** To report prospectively the prenatal diagnosis, management and outcome of 14 cases of unilateral ventriculomegaly.

**Methods** Fourteen fetuses were diagnosed as having one ventricle of  $\geq 10$  mm, as measured at the level of the atrium.

**Results** In ten cases, the scan showed mild unilateral ventriculomegaly with an atrium width between 11 and 13 mm and this remained stable up to term. Eight of these fetuses had a magnetic resonance imaging scan in utero between 32 and 34 weeks of gestation which confirmed the diagnosis of mild ventriculomegaly without other brain abnormalities and showed a normal cortical mantle. No obvious cause was found and the outcome was normal in all cases. In four cases, the unilateral ventriculomegaly evolved rapidly with an atrium width up to 20–25 mm. Causes included atresia of the foramen of Monro, toxoplasmosis, brain atrophy and Weaver syndrome. Three underwent termination of pregnancy and the postmortem examination confirmed the diagnosis. The baby with brain atrophy and schizencephaly had a ventriculoperitoneal shunt placed at 1 month of age and has severe developmental delay at 9 months.

**Conclusion** The prognosis of unilateral ventriculomegaly is uncertain. Examination of both ventricles during the anomaly scan should be performed, as should ultrasound follow-up of these cases up to the end of the third trimester. Fetuses with an isolated, mild, stable unilateral ventriculomegaly seem to have a favourable neurological outcome. However, fetuses with rapidly evolving unilateral ventriculomegaly or cases associated with other brain abnormalities may have a poor neurological outcome.

## INTRODUCTION

Unilateral hydrocephalus has been defined as the progressive dilatation of one lateral ventricle due to an abnormal circulation of cerebrospinal fluid (CSF)<sup>1</sup>. Obstruction of the foramen of Monro is the most common cause of this rare entity, the prevalence of which is unknown. The foramen of Monro can be obstructed by congenital atresia<sup>2,3</sup>, morphological obstruction due to hemorrhage<sup>4</sup>, neoplasm<sup>5</sup>, gliomatous<sup>6</sup> or vascular anomalies<sup>7</sup>, and physical obstruction due to infection<sup>8</sup> or trauma. Functional obstruction can develop after ventriculostomy due to one-way valve action at the foramen of Monro<sup>1</sup>. The term ventriculomegaly is used in preference to hydrocephalus because ventriculomegaly more accurately describes the abnormality found on antenatal ultrasonography<sup>9</sup>. On the basis of a small series it seems that unilateral ventriculomegaly is a separate entity from bilateral ventriculomegaly, with a lower risk of perinatal mortality and morbidity; fewer cases are associated with central nervous system or other anomalies<sup>1,10–12</sup>. However, the prognosis depends largely on the severity of the dilatation with therapeutic options ranging from expectant management to surgical ventriculostomy.

Prenatal sonography has been shown to diagnose congenital bilateral ventriculomegaly reliably; however, only a few cases of prenatal diagnosis of unilateral ventriculomegaly have been reported<sup>10–18</sup>.

## METHODS

This prospective study included 14 fetuses in which unilateral ventriculomegaly was diagnosed during a second- or third-trimester ultrasound examination between January 1995 and July 1998. The study population included pregnant women at low risk of fetal congenital abnormalities at

the time of the routine scan together with patients referred for suspicion of fetal unilateral ventriculomegaly. All fetuses presenting with unilateral ventriculomegaly, with or without other brain abnormalities, were included.

Brain examination was performed by obtaining an axial plane passing through the frontal horns and atria of the lateral ventricles. In addition, coronal, sagittal and parasagittal planes were obtained to visualize both lateral ventricles and to enable a more detailed evaluation of the fetal brain to be made. In cases in which the head was sufficiently low in the maternal pelvis, a transvaginal scan was also undertaken. Unilateral ventriculomegaly of the lateral ventricles was defined when only one ventricle measured  $\geq 10$  mm at the level of the atrium. We measured the atrial width directly on the screen using the electronic calipers to obtain the inner-to-inner width. The calipers were positioned on the luminal margins of the ventricular wall at the junction of the ventricular wall and the ventricular lumen, as previously described<sup>19,20</sup>, perpendicular to the long axis of the ventricle. IgG and IgM antibodies against toxoplasma, rubella, cytomegalovirus and herpes (TORCH) were assayed in maternal serum in all cases. Karyotyping was discussed in all cases and was performed according to the patients' request and the cause of and prognosis for the ventriculomegaly. In eight cases, magnetic resonance imaging (MRI) was performed *in utero* between 32 and 34 weeks in order to improve the visualization of the fetal brain. Monthly follow-up ultrasound examinations were performed if the ventriculomegaly was stable, and weekly if the dilatation was evolving. Ultrasound and MRI examinations were reviewed during the pregnancy in all cases by a pediatric neuroradiologist. Transfontanellar ultrasound scans were performed on all neonates after delivery. Pediatric follow-up was available in all cases.

## RESULTS

Fourteen fetuses were diagnosed prenatally as having unilateral ventriculomegaly (Figures 1 and 2). Ten fetuses had an isolated, mild and stable dilatation between 11 and 13 mm up to term. Eight of these fetuses had a brain MRI scan *in utero*, performed between 32 and 34 weeks of gestation, which confirmed the diagnosis of mild ventriculomegaly without other brain abnormalities and showed a normal cortical mantle. No obvious cause was found and all these babies show normal development at 3–48 months of age. Four fetuses had mild to moderate unilateral ventriculomegaly associated with brain abnormalities or other malformations (Table 1).

### Case 1

A 35-year-old multiparous woman was diagnosed as having a 12-week fetus with a markedly increased nuchal translucency and generalized skin edema, but normal karyotype. Serial ultrasound scans revealed early fetal overgrowth from 16 weeks onwards and suggested additional brain abnormalities including mild, stable unilateral ventriculomegaly and a cyst in the cavum septum pellucidum.

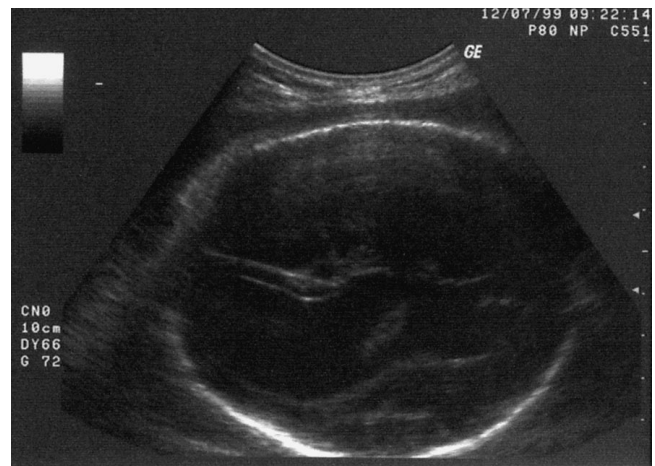


Figure 1 Axial view of the brain of a 24-week fetus showing unilateral ventriculomegaly

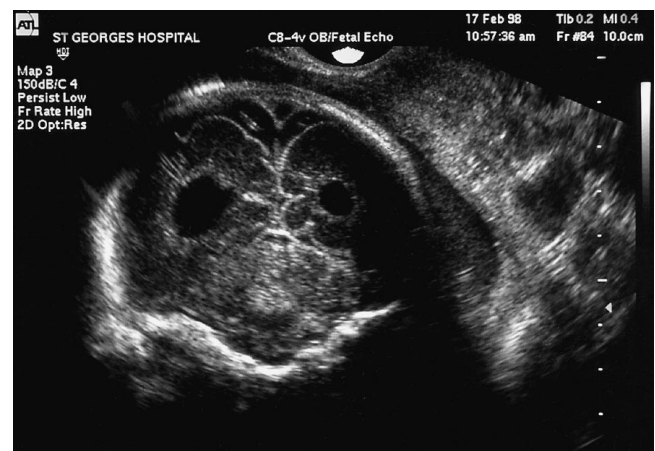


Figure 2 Coronal view of the brain of a 24-week fetus showing unilateral ventriculomegaly

The diagnosis of genetic overgrowth syndrome was suggested on the basis of excessive fetal growth (+ 4 SD above the mean), advanced bone age (as assessed on intra-uterine X-ray) and characteristic facial features. The pregnancy was terminated at the parents' request at 34 weeks of gestation. Postmortem examination confirmed the prenatal diagnosis of Weaver syndrome<sup>21</sup>.

### Case 2

A 36-year-old primigravida was referred to our center at 31 weeks of gestation for the management of unilateral ventriculomegaly. The scan showed a left atrium width of 16 mm with ipsilateral hyperechogenicity of the choroid plexus (Figure 3). One week later, the left ventriculomegaly was surrounded by hyperechogenic areas within the parenchyma at the junction of the lateral ventricle and the septum pellucidum and in the basal ganglia. The choroid plexus on the left side was very thin, heterogeneous and hyperechogenic. A maternal blood sample and second amniocentesis confirmed the diagnosis of congenital toxoplasmosis. Termination of pregnancy was performed at 33 weeks of gestation.

**Case 3**

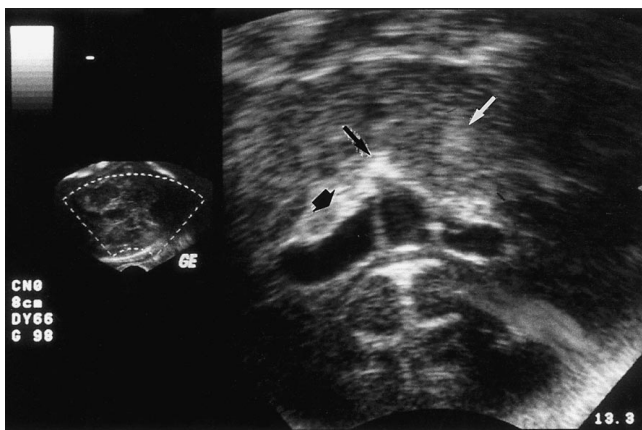
A 19-year-old primigravida was diagnosed as having a fetus with unilateral ventriculomegaly with an atrium width of 11 mm at 21 weeks. No other cranial or extracranial abnormalities were detected. One month later,

the scan showed mild stable ventriculomegaly but the choroid plexus was bright and irregular in shape, suggesting previous bleeding. The MRI and ultrasound scans performed at 30 weeks showed severe damage to the wall of the ventricle. The cortex was thin and there was evidence of posterior schizencephaly (Figures 4 and 5). The couple

**Table 1** Clinical characteristics of 14 fetuses with prenatally diagnosed unilateral ventriculomegaly

Case	Sex	Maternal age (years)	GA at diagnosis (weeks)	GA at delivery (weeks)	Atrium size (mm)	Normal contra-lateral ventricle Side	Evolution	MRI	Causes	Birth weight (g)	Outcome	Follow-up (month)
1	F	35	16	34	13	R 9	stable	yes	Weaver syndrome	3450	termination	
2	M	37	31	33	16–20	L 8	progressive	no	toxoplasmosis	2150	termination	
3	M	19	21	40	11–13	R 7	progressive	yes	brain atrophy	3500	severe developmental delay	12
4	M	28	21	31	13–24	L 7	progressive	yes	agenesis of foramen of Monro with porencephaly	1900	termination	
5	M	24	31	40	12	R 7	stable	yes	unknown	3860	normal development	16
6	M	24	21	41	11	R 7.5	stable	no	unknown	4820	normal development	9
7	M	37	24	41	13	L 9	stable	no	unknown	4640	normal development	6
8	F	27	21	40	12	L 7	stable	yes	unknown	3220	normal development	12
9	M	31	19	40	12	L 7.5	stable	yes	unknown	3680	normal development	14
10	M	35	31	41	12	L 7	stable	yes	unknown	3980	normal development	3
11	M	22	32	41	12	R 8.5	stable	yes	unknown	3880	normal development	48
12	M	25	32	41	12	L 9	stable	yes	unknown	4060	normal development	36
13	M	27	32	40	13	L 7.5	stable	yes	unknown	3880	normal development	12
14	F	35	32	39	11	R 8	stable	yes	unknown	4000	normal development	12

GA, gestational age; MRI, magnetic resonance imaging; F, female; M, male; R, right; L, left



**Figure 3** Coronal view of the brain at 31 weeks showing unilateral ventriculomegaly with hyperechogenicity of the choroid plexus (large arrow) and the thalami (thin arrows) in relation to toxoplasmosis



**Figure 4** Parasagittal views of the brain at 25 weeks showing rupture of the cortical mantle (arrow) and schizencephaly



**Figure 5** Sagittal magnetic resonance image showing increased pericerebral space and atrophy of the cortical mantle in the posterior part of the brain (large arrow) and schizencephaly (thin arrow). The anterior cortical mantle appears to have normal thickness

was informed that the prognosis was very poor, but they decided to continue with the pregnancy. At 9 months of age, the child has severe developmental delay.

#### Case 4

A 28-year-old primigravida was diagnosed at the time of the anomaly scan as having a fetus with unilateral ventriculomegaly and an atrium width of 13 mm. One month later, the scan showed moderate unilateral ventriculomegaly. At 30 weeks, the ventriculomegaly was marked, with severe thinning of the cortex on the affected side and poor definition of the lateral ventricular wall. The MRI scan confirmed the diagnosis of marked unilateral ventriculomegaly and suggested porencephaly. A termination was performed at 31 weeks of gestation. Agenesis of the foramen of Monro with porencephaly was diagnosed at the postmortem examination.

## DISCUSSION

Demonstration of the fetal ventricles is important, since ventriculomegaly is one of the most common abnormalities detected by prenatal sonography. Different ventricular measurements have been used, including the ratio of the lateral ventricle to the hemispheric widths, measurement of the frontal and occipital horns, the size of the atrium and the position of the choroid angle<sup>22</sup>. Regardless of the method chosen to evaluate the width of the ventricle, the axial plane does not allow both ventricles to be imaged simultaneously owing to the presence of reverberation artifacts masking much of the hemisphere proximal to the

transducer. This can, however, be visualized by angling the ultrasound beam through the anterolateral fontanelle<sup>23</sup>, changing the maternal position or waiting until the fetal movement enables adequate visualization. Alternatively, a coronal plane of the fetal head can be obtained by combining transabdominal and transvaginal ultrasound examinations if the head is low in the maternal pelvis. We believe that both ventricles should be visualized in all routine ultrasound examinations if unilateral ventriculomegaly is to be excluded.

Inherent in all measurements is the need to identify the lateral wall of the lateral ventricle. Although this wall can be identified in virtually all patients if it is specifically sought<sup>24</sup>, this may be problematic in the second trimester, owing to the choroid plexuses, which are large and echogenic. In addition, they fill the entire lateral ventricle posterior to the foramen of Monro, tending to mask the ipsilateral ventricle wall. These factors, together with the hypoechogenicity of the surrounding brain tissue, can cause ventriculomegaly to be misinterpreted. This artifact disappears when coronal images are obtained. Among all landmarks used to evaluate ventricular size, the atrium of the lateral ventricle seems to be the most pertinent site<sup>25-28</sup>. Indeed, the diameter of the ventricular atrium remains stable throughout the second and third trimesters and its appearance does not change with gestation. Moreover, the atrium is easily recognized, owing to the presence of the glomus of the choroid plexus, which is among the most conspicuous intracranial landmarks. The walls of the atrium are typically perpendicular to the ultrasound beam in the axial plane and this can be identified in virtually 100% of the fetuses<sup>24</sup>.

In our review of 38 published cases of prenatal diagnosis of unilateral hydrocephalus or unilateral ventriculomegaly, a cause was identified in 11 cases including three cases with a poor neurological outcome (Table 2). Twenty-seven cases were considered to be isolated, unexplained, borderline unilateral ventriculomegaly with a good neurological outcome (Table 2). Prognostic factors are difficult to identify and mild dilatation at the time of the diagnosis cannot be taken as reassuring. In all 11 published cases in which the cause was known, information on prenatal scanning was unavailable. In our series (Table 1), three fetuses with mild ventriculomegaly at the time of diagnosis (11–13 mm) had a poor outcome. Following initial assessment by ultrasound, infection screening and follow-up, including serial ultrasound examinations, MRI and multidisciplinary discussions, it appears that unilateral, isolated, borderline and stable ventriculomegaly up to term is likely to represent a normal anatomical variation of the fetal brain. The upper limit of the normal range for the lateral ventricles in an axial plane is usually accepted as 10 mm<sup>21,26,28-30</sup>. These investigators suggested that 10 mm was a value approximately four standard deviations above the mean and would therefore be an acceptable upper limit, although this limit has not been universally applied to investigate ventricular abnormalities. A large prospective study suggested that the cut-off level currently used to define normal atrial size was too low and suggested 12 mm as the upper limit<sup>31</sup>.

**Table 2** Summary of 38 reported cases of prenatally diagnosed unilateral hydrocephalus

Case	Etiology	Treatment	Outcome	Reference
1	membranous occlusion of the foramen of Monro (multiple malformations)		stillborn	11
2	atresia of the foramen of Monro	V-P shunt	left hemiparesia; global developmental delay	15
3	congenital obstruction of the foramen of Monro	V-P shunt	normal cognitive development	12
4	atresia of the foramen of Monro	V-P shunt	normal cognitive development	11
5	atresia of the foramen of Monro	V-P shunt	normal cognitive development	11
6	atresia of the foramen of Monro with porencephaly	V-P shunt	severe developmental delay	11
7	obstruction of the foramen of Monro due to intraventricular hemorrhage	V-P shunt	normal cognitive development	11
8	underlying brain dysplasia with holoprosencephaly	V-P shunt	moderate developmental delay	11
9	unknown	V-P shunt	normal cognitive development	11
10	obstruction of the foramen of Monro by a frontoethmoidal encephalocele	craniotomy	normal cognitive development	14
11	obstruction of the foramen of Monro by a septum with check-valve effect	no treatment	normal cognitive development	16
12–38	unknown	no treatment	$n = 25$ , normal cognitive development; $n = 1$ epilepsy; $n = 1$ termination of pregnancy (normal postmortem examination)	17

V-P, Ventriculoperitoneal

Moreover, a large retrospective study showed that male fetuses had a slightly larger atrial size<sup>32</sup>. We found that eight out of ten fetuses with isolated unilateral ventriculomegaly had a birth weight of  $\geq 90$ th centile, and eight newborns were boys. These findings raise the question of whether different normal ranges should be established according to birth weight and sex.

## CONCLUSION

Prenatal detection of unilateral ventriculomegaly is difficult, because common sonographic artifacts frequently obscure the ventricle closer to the transducer. Moreover, the prognosis of unilateral ventriculomegaly can be uncertain. This calls for a systematic examination of both ventricles during a detailed anomaly scan using axial, coronal, sagittal and parasagittal planes, with focused high-resolution sonographic transducers. Prenatal diagnosis of unilateral ventriculomegaly calls for detailed assessment and follow-up by serial ultrasound examinations up to term. A large proportion of mild and stable cases in large-for-dates and/or male fetuses are likely to represent a variation of the normal fetal anatomy.

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