ORIGINAL ARTICLE

Ventriculomegaly and its Association with CNS Anomalies: Can Ventriculomegaly in CNS Anomaly be Compared with Axis Deviation in Cardiac Anomaly? A Retrospective Study

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Abstract

Aim: The purpose of this retrospective study was to examine the association of ventriculomegaly in cases of CNS anomaly and to study the sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV) of ventriculomegaly in predicting CNS anomaly. We have also tried to compare the predictive value of ventriculomegaly in predicting CNS anomaly to cardiac axis deviation in predicting cardiac anomaly.

Materials and methods: Over a period of 5 years, all cases of CNS anomalies with or without ventriculomegaly with gestational age more than 14 weeks were reviewed for the size of lateral ventricles, other CNS findings, extra-CNS structural anomalies. Ventricle size up to 7.0 mm was considered normal, the size from 7.1 to 10.0 mm was considered as prominent ventricle, from 10.1 to 14.9 mm was considered as mild ventriculomegaly and the size of ≥15 mm was considered severe ventriculomegaly.

Results: In total, there were 206 CNS anomalies detected in 165 patients. The commonest CNS anomaly detected was agenesis of corpus callosum (ACC) and partial agenesis of corpus callosum (PACC). Our data show that although ventriculomegaly is a strong marker for suspecting intracranial structural abnormality, anomalies of posterior fossa, PACC, and intracranial cysts may be present with normal-sized ventricles.

Conclusion: From our study, we can conclude that ventriculomegaly does not predict the presence of structural CNS abnormality, and a normal size ventricle rules does not rule out the presence of structural CNS anomaly. Ventriculomegaly is not a strong marker in CNS anomaly like cardiac axis deviation in cardiac anomaly.

Keywords: Agenesis of corpus callosum, CNS anomaly, Lateral ventricle, Posterior fossa anomaly, Prominent ventricle, Ventriculomegaly.

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Introduction

Ventriculomegaly (VM) is an enlargement of lateral cerebral ventricle caused by an excess of cerebrospinal fluid. Fetal ventriculomegaly is defined as a ventricular atrium which measures 10 mm or greater at any gestation.[1]

The incidence of VM ranges from 0.3 to 1.5% live births.[2] Unilateral VM occurs in 60% of the cases, leaving 40% bilateral VM. There is a male predominance (70%). Ventriculomegaly may result from obstructive malformations, destructive lesions, and abnormal development of the brain. The diagnosis is made with the measurement of the lateral ventricle in a strict axial plane on ultrasonography. In mild VM, structural anomalies range from 10 to 76%. However, even in apparently isolated VM, malformations are found in 13% of the cases after birth. Chromosomal anomalies are detected in more than 15% of the cases when VM is associated with other fetal anomalies.

Aim

The purpose of this retrospective study was to examine the association of ventriculomegaly in the cases of CNS anomaly and to study the sensitivity, specificity, positive, and negative predictive value (NPV) of ventriculomegaly in predicting the CNS anomaly. We have also tried to compare the predictive value of ventriculomegaly in predicting CNS anomaly to cardiac axis deviation in predicting cardiac anomaly.

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Materials and Methods

This is a retrospective study of all cases of CNS anomalies presenting in a tertiary fetal medicine center between January 1, 2015 and December 31, 2019 (5 Years). Ultrasound scanning for all selected cases was performed by using Voluson E10 Machine and Volume Probe.

Cases were selected based on the following inclusion and exclusion criteria.

Inclusion Criteria

All cases of CNS anomalies with or without ventriculomegaly with gestational age (GA) more than 14 weeks were selected.

Exclusion Criteria

All cases of open neural tube defects were excluded from the study.

Cases were reviewed for GA at presentation, size of lateral ventricles (both right and left), other CNS findings, structural anomalies other than CNS anomaly. Detailed ultrasound examination with fetal echocardiography and fetal neurosonogram [preferably transvaginal ultrasound (TVS) whenever the presentation was cephalic] was done to exclude other structural abnormalities.

The cases were categorized based on the size of lateral ventricles and type of CNS and extra-CNS anomalies and gestational age. All ventricular measurements were obtained with transabdominal US (UltraSound) probe according to the International Society of Ultrasound in Obstetrics and Gynecology guidelines. The atrial width at the level of the glomus of the choroid plexus was measured by placing calipers inside the echoes generated by medial and lateral ventricular walls in transventricular plane. Both lateral ventricles were measured in order to characterize unilateral and bilateral dilation as well as symmetry between ventricles. Ventricle size up to 7.0 mm was considered normal, ventricle size from 7.1 to 10.0 mm was considered as prominent ventricle, and the size from 10.1 to 14.9 mm was considered mild ventriculomegaly and ventricle size of ≥15 mm was considered as severe ventriculomegaly. Asymmetry was defined as a difference of ≥2 mm between the two lateral ventricles.

The cases were classified into four groups as follows: group I: Cases with no other CNS or extra-CNS findings other than ventriculomegaly were included in the isolated VM group; group II: Ventriculomegaly with CNS anomalies only; group III: Ventriculomegaly with extra-CNS anomalies only; and group IV: Ventriculomegaly with CNS + extra-CNS anomalies.

Results

Over the period of 5 years, 8224 pregnant women were screened. 1642 scans were abnormal with the incidence of 19.95%. Total of 240 CNS anomalies were detected with 2.91% as the incidence of CNS anomaly. About 59 cases (0.71%) having open neural tube defect were excluded from the study. Out of 181 cases of CNS anomalies, 165 cases fulfilled the inclusion criteria and were included in the study. Few patients had more than 1 CNS anomaly, whereas some had isolated ventriculomegaly. In total, there were 206 CNS anomalies detected in 165 patients.

Table 1 presents the istribution of cases with CNS anomalies based on the size of ventricles.

There were 57 cases of CNS anomaly with normal-sized ventricles, 41 cases with prominent ventricles, 58 cases with ventriculomegaly, and 9 cases where ventricular size was not measurable. Such cases were of alobar holoprosencephaly where ventricular size was not measured due to the presence of monoventricle, and others were of less than 16 weeks gestation, where measurement of ventricular size was not possible.

Table 2 depicts the different types of CNS anomalies in our study.

Table 1: Distribution of cases with CNS anomalies based on the size of ventricles

Size of ventricles	*No of cases, N = 165*	*Percentage (%)*
Upto 7 mm (Normal)	57	34.5
7.1–10 mm (Prominent Ventricles)	41	24.8
≥10.1 mm (Ventriculomegaly)	58	35.2
Size not measurable	09	5.5
Total	165	

The most common CNS anomaly detected was agenesis of corpus callosum (ACC) and partial agenesis of corpus callosum (PACC) with 28 cases of each, followed by 22 cases of cystic lesions of brain (which included arachnoid cyst, inter-hemispheric cyst, chronic venous insufficiency (CVI) cyst, frontal horn cyst, and periventricular cysts).

Figure 1 shows the cases of ventriculomegaly and its categorization.

There are total 58 cases of ventriculomegaly, out of which 15 are isolated ventriculomegaly, that is, not associated with any other anomaly. Out of these 15 cases, 10 were mild ventriculomegaly and 5 were severe ventriculomegaly. There were 26 cases of ventriculomegaly, which were associated with CNS anomalies only, out of which 12 were mild and 14 were severe ventriculomegaly. There were 17 cases which had CNS + Extra-CNS anomalies, out of which 10 were mild and 7 were severe ventriculomegaly.

Figure 2 shows the incidence of prominent ventricles and its categorization.

There are 41 cases of prominent ventricles, out of which 7 were Isolated. About 24 cases of prominent ventricles were associated with CNS anomalies alone, 2 cases were associated with extra-CNS anomalies alone, whereas 8 cases were associated with CNS + Extra-CNS anomalies.

Table 3 presents the CNS anomalies that were associated with normal-sized lateral ventricles.

There were totally 57 cases, out of 165 cases of CNS anomaly which had normal-sized ventricles. The cases which had normal ventricles have been listed in the Table 3 along with the gestational age.

Table 4 depicts the categorization of the anomalies according to the size of the ventricles.

As seen in Table 4, 67.8% of ACC cases had ventriculomegaly, whereas 33.2% had normal or prominent ventricles. Similarly, only 17.8% cases of PACC had ventriculomegaly and 82.2% had normal or prominent ventricles. About 40.9% of intracranial cyst cases had ventriculomegaly. Only 15.7% cases of Dandy-Walker malformation (DWM) had ventriculomegaly while the majority cases of DWM 84.3% had normal size ventricle. About 38.8% of hypoplastic cerebellum had ventriculomegaly and 68.7% of lobar and semilobar holoprosencephaly had ventriculomegaly.

Postnatal follow-up was taken in cases of isolated prominent ventricles and isolated ventriculomegaly. Table 5 presents the postnatal follow-up in cases of isolated prominent ventricles and isolated VM.

To find out the statistics of ventriculomegaly in predicting CNS anomalies, we used 2 × 2 table and used PPV and NPV calculator to find out the values. Table 6 presents the statistics of ventriculomegaly in predicting CNS anomalies. The sensitivity and

Table 2: List of CNS anomalies according to gestational age

Anomalies	*Upto 20 weeks N = 37*	*20.1–30 weeks N = 144*	*30.1–40 weeks N = 26*	*Mean GA at diagnosis (weeks)*	*Total N = 207*
Agenesis of corpus callosum (ACC)	3	21	4	24 + 1	28
Partial agenesis of corpus callosum (PACC)	4	23	1	23 + 3	28
Cystic lesions of brain	5	14	3	25 + 2	22
Dandy Walker malformation	7	12	0	20 + 3	19
Hypoplastic cerebellum	1	13	4	25 + 3	18
Holoprosencephaly (lobar + alobar)	4 (3 + 1)	11 (9 + 2)	1 (1 + 0)	22 + 0	16 (13 + 3)
Isolated ventriculomegaly	0	12	3	26 + 0	15
Mega cisterna magna	0	6	4	28 + 4	10
Isolated prominent ventricles	3	4	0	21 + 3	7
Isolated inferior vermian hypoplasia (IIVH)	0	7	1	26 + 0	8
Encephalocoele	5	3	0	18 + 1	8
Microcephaly	2	3	1	23 + 4	6
Septo-optic dysplasia + agenesis of septum pellucidum	1	5	0	23 + 3	6
Blake pouch cyst	1	4	0	22 + 1	5
Intracranial hemorrhage	1	2	1	26 + 2	4
Schizencephaly	0	1	1	27 + 5	2
Intra cerebellar infarct	0	1	0	26 + 4	1
Vein of Galen malformation	0	0	1	35 + 1	1
Rhombencephalosynapsis	0	1	0	23 + 6	1
Choroid plexus papilloma	0	0	1	32 + 5	1
Lipoma in skull	0	1	0	22 + 3	1
Total					207

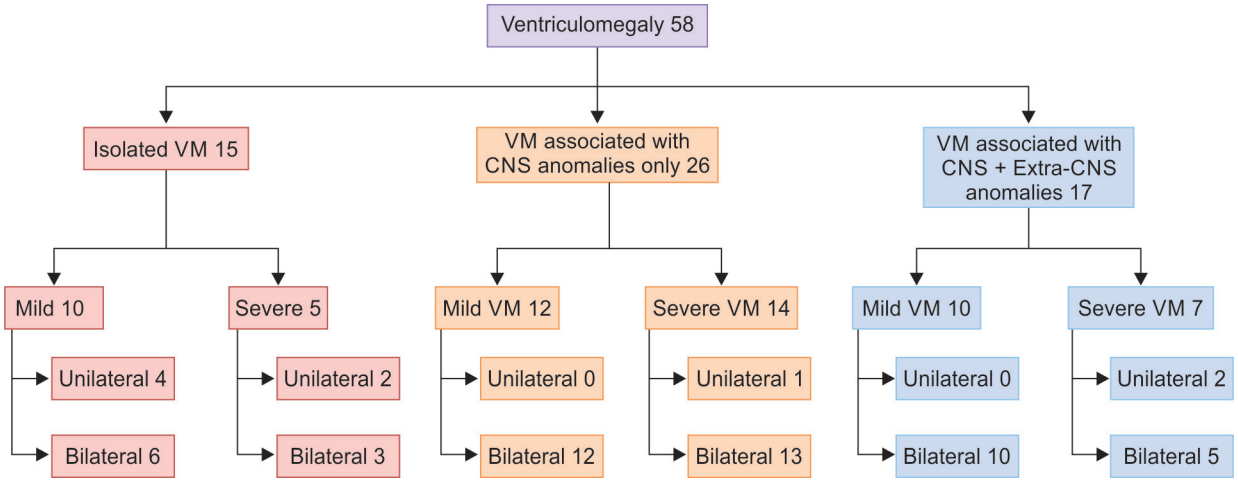

Fig. 1: Ventriculomegaly and its categorization

specificity of ventriculomegaly in predicting the CNS anomalies is 43% and 73.21%, respectively, whereas the positive predictive value and negative predictive value of ventriculomegaly in predicting CNS anomalies is 74.14% and 41.84%, respectively.

Discussion

In this study, we analyzed a large number of fetuses diagnosed with prominent ventricles and ventriculomegaly. The data shown in this study help in establishing the correlation between lateral ventricle width and CNS anomalies.

As seen in Table 2, the maximum cases of CNS anomalies which we diagnosed were of ACC and PACC.

There were totally 57 cases, out of 165 cases of CNS anomaly which had normal-sized ventricles, 13 cases out of which were of PACC, all of whom were diagnosed between 20 and 30 weeks of gestation.

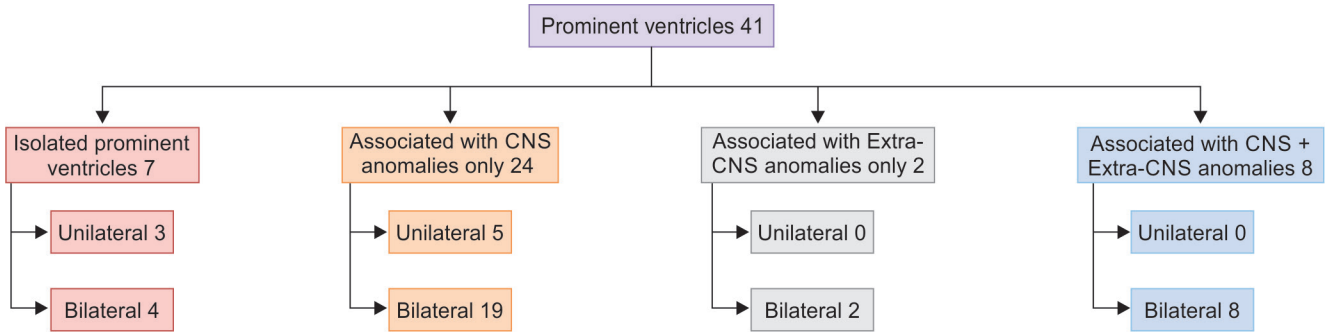

Fig. 2: Prominent ventricles and its categorization

Table 3: CNS anomalies associated with normal ventricles according to gestational age

CNS anomaly	*Total*	*< 20 weeks*	*20.1–30 weeks*	*>30 weeks*
PACC	13	–	13	–
Dandy Walker malformation	9	3	6	–
Mega cisterna magna	5	–	4	1
Intra cranial cysts	5	2	2	1
ACC	5	–	5	–
Blake pouch cyst	4	1	3	–
Hypoplastic cerebellum	4	–	4	–
Encephalocoele	3	–	3	–
IIVH	2	–	2	–
SOD + agenesis of septum pellucidum	2	–	2	–
Lipoma in skull	1	–	1	–
Rhombencephalosynapsis	1	–	1	–
Vein of Galen malformation	1	–	–	1
Intracranial hemorrhage	1	1	–	–
Microcephaly	1	–	1	–
Total	57			

Table 4: Categorization of the anomalies according to the size of the ventricles: (Excluding the cases of isolated ventriculomegaly and isolated prominent ventricles)

Anomaly	*Total*	*Normal ventricles*	*Prominent ventricles*	*Ventriculomegaly*	*Size not measurable*	*Cases with normal + Prominent ventricles/Cases with ventriculomegaly*
ACC	28	5	4	19 (67.8%)		9/19
PACC	28	13	10	5 (17.8%)		23/5
Intracranial cysts	22	8	5	9 (40.9%)		13/9
Dandy Walker malformation	19	10	1	3 (15.7%)	5	11/3
Hypoplastic cerebellum	18	9	2	7 (38.8%)		11/7
Holoprosencephaly (lobar + alobar)	16 (13+3)	– (0+0)	2 (2+0)	11 (68.7%) (11+0)	3 (0+3)	2/11
Mega cisterna magna	10	5	2	3 (30%)		7/3
Encephalocoele	8	3	1	1 (12.5%)	3	4/1
IIVH	8	5	2	1 (12.5%)	–	7/1
Microcephaly	6	3	1	1 (16.6%)	1	4/1
SOD + agenesis of septum pellucidum	6	2	4	(0%)		6/0
Blake pouch cyst	5	5	–	–(0%)	–	5/0

Intracranial hemorrhage	4	1	1	2 (50%)		2/2
Schizencephaly	2		1	1 (50%)		1/1
Vein of Galen malformation	1	1		(0%)		1/0
CP papilloma	1			1 (100%)		0/1
Lipoma in skull	1	1		(0%)		1/0
Rhombencephalosynapsis	1	1		(0%)		1/0
Cerebellar infarct	1		1	(0%)		1/0
Total	185				12	109/64

Table 5: Postnatal follow-up of isolated ventriculomegaly and isolated prominent ventricles

	Mild VM (N = 10)	*Severe VM (N = 5)*	*Prominent ventricles (N = 7)*
Normal on follow-up	4	1	5
Died after birth	2	1	–
Terminated pregnancy	2	3	1
Lost to follow-up	2	–	1

Compared with PACC, ACC had more association with ventriculomegaly (67.8% of ACC cases had ventriculomegaly compared with00 only 17.8% of PACC cases). We can see that majority of the cases of PACC (82.2%) had normal-sized ventricles at the time of diagnosis.

Other anomaly after PACC which was associated with normal-sized ventricles was posterior fossa anomalies. Out of 62 cases of posterior fossa anomalies (which includes DWM, Blake pouch cyst, mega cisterna magna, hypoplastic cerebellum, isolated inferior vermian hypoplasia (IIVH), rhombencephalosynapsis and cerebellar infarct). Only 14 cases (22%) had ventriculomegaly.

About 68.7% cases of lobar holoprosencephaly had associated ventriculomegaly at the time of diagnosis. Among the cases of intracranial cysts, 40.9% cases had ventriculomegaly.

The positive predictive value and negative predictive value of ventriculomegaly in predicting CNS anomalies is 74.14% and 41.84%, respectively. We can see from the above values that ventriculomegaly is not a strong marker in predicting CNS anomalies, nor does a normal-sized ventricle rules out the presence of CNS anomaly. These findings are contrary to previous studies, which say that the rate of minor and major findings increases with each millimeter increase in ventricle width.[3]

One of the advantages of this study is that it included a large number of fetal scans, enabling us to examine in-depth the association between ventriculomegaly and CNS findings.

Abnormal cardiac axis is almost always associated with cardiac anomaly or intra-thoracic abnormality.[4] Abnormal cardiac axis is the first pointer toward suspected cardiac anomaly even in the first trimester scan.[5] However, our data show that although ventriculomegaly is a strong marker for suspecting intracranial structural abnormality, anomalies like posterior fossa anomalies, PACC, and intracranial cysts may be present with normal-sized ventricles. About 34.5% of CNS anomaly had normal ventricles, and only 35.2% cases had ventriculomegaly. Normal-sized ventricles do not rule out CNS anomaly and a thorough scan of the fetal head is proposed even in cases of normal ventricles to avoid missing out on an important anomaly.

Table 6: Ventriculomegaly in predicting CNS Anomalies

Sensitivity	43%
Specificity	73.21%
Positive predictive value	74.14%
Negative predictive value	41.84%

Conclusion

Excluding cases of isolated prominent ventricles and isolated ventriculomegaly, there were a total of 185 anomalies, out of which 12 anomalies had non-measurable ventricles. Out of remaining 173 anomalies, 109 had normal + prominent ventricles and only 64 cases had ventriculomegaly (34%). Certain anomalies like posterior fossa anomalies, PACC and intra cranial cysts had normal-sized ventricles. Ventriculomegaly was present in 35.2% of the cases of CNS anomaly and 34.5% cases of CNS anomaly had normal-sized ventricles. In cardiac evaluation, an abnormal cardiac axis is the first indicator toward cardiac anomaly. The PPV of abnormal axis in cardiac anomaly is very high. Whereas in our study, the PPV and NPV of ventriculomegaly in predicting the CNS anomalies is 74.14% and 41.84%, respectively. Also, sensitivity and specificity of ventriculomegaly in detecting CNS anomalies is 43% and 73.21% only. From our study, we can conclude that ventriculomegaly does not predict the presence of structural CNS abnormality nor a normal-sized ventricle rules out the presence of structural CNS anomaly. A detailed and thorough evaluation of fetal brain is necessary even in the presence of normal-sized lateral ventricles so as to avoid missing out any CNS anomaly.

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