# The Dandy–Walker variant: a case series of 24 pediatric patients and evaluation of associated anomalies, incidence of hydrocephalus, and developmental outcomes

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Object. The Dandy–Walker complex is a continuum of aberrant development of the posterior fossa that has been associated with multiple congenital anomalies, radiographic abnormalities, and developmental delay. The Dandy–Walker variant (DWV) is a unique entity believed to represent a milder form of the complex, and is characterized by a specific constellation of radiographic findings. In this retrospective case series, the authors report the association of the DWV with other congenital anomalies, the associated radiographic findings linked with DWV, and the developmental outcome in this population.

*Methods*. The charts and radiographs of 10 male and 14 female patients treated between 2000 and 2006 were examined. The patients' mean gestational age was 35.6 weeks (range 23–41 weeks), and the mean follow-up period was 5.1 years (range 1 month–15 years).

Results. Three patients died. Associated anomalies included cardiac (41.7%), neurological (33.3%), gastrointestinal (20.8%), orthopedic (12.5%), and genitourinary (12.5%) abnormalities. Less common were pulmonary and psychiatric findings. Developmental delay was identified in 11 of the 21 patients for whom follow-up was available. Five of 6 patients with isolated DWV had a normal developmental course. Radiographic findings associated with DWV included corpus callosum dysgenesis in 20.8%, ventricular enlargement in 29%, and vermian rotation in 8.3%. Shunts were placed in 4 of 7 patients with ventriculomegaly. Using the two-tailed Pearson correlation, the authors determined that developmental outcome was solely affected by neurological deficits and that ventricular enlargement predicted the need for shunt placement.

Conclusions. The DWV was associated with both extra- and intracranial anomalies. Associated radiographic abnormalities including ventriculomegaly were observed. Hydrocephalus requiring cerebrospinal fluid diversion may be indicated. Isolated DWV was associated with a good developmental outcome. (DOI: 10.3171/PED/2008/2/9/194)

KEY WORDS • Dandy-Walker complex • Dandy-Walker malformation • Dandy-Walker variant • hydrocephalus • posterior fossa anomaly • posterior fossa cystic lesion

YSTIC anomalies of the posterior fossa in children can be grouped along a continuum that has become collectively known as the Dandy–Walker complex. 3,5,6,8, 10,11,15,16,19-21,27,29-31,34,35,37 The severity of the diagnosis is based on the size of the posterior fossa, the presence of cystic dilatation of the fourth ventricle, and the degree of vermian hypoplasia. Simple enlargement of the posterior fossa with an enlarged foramen magnum and associated cisterna magna, which is known as "mega cisterna magna," is usually a

Abbreviations used in this paper: CNS = central nervous system; CSF = cerebrospinal fluid; DWM = Dandy–Walker malformation; DWV = Dandy–Walker variant.

benign finding when not associated with other congenital anomalies. Retrocerebellar arachnoid cysts can be differentiated from the DWM and DWV in that there is no communication between the cyst and the fourth ventricle.

At the other end of the spectrum is the well-known entity DWM, which is characterized by the presence of a large posterior fossa cyst with open communication between the fourth ventricle, absent or severely atretic inferior vermis, and enlargement of the posterior fossa with elevation of the confluence of sinuses, lateral sinuses, and tentorium. <sup>3,6,7,10,15,16,18,19,21,27,30</sup> The association of DWM with intra- and extracranial congenital anomalies, developmental delay, and hydrocephalus is well established. The DWV is believed to be

a less severe anomaly and is characterized by mild vermian hypoplasia and a normal-sized posterior fossa with a small cystic cavity that communicates with the fourth ventricle. 8.15,16,20 The association of DWV with other congenital anomalies, radiographic abnormalities, incidence of hydrocephalus, and developmental outcomes is largely limited to case reports. The purpose of this retrospective case series was to evaluate the association of DWV with other congenital anomalies, describe other associated radiographic characteristics, and evaluate developmental outcomes.

#### Methods

A retrospective Internal Review Board-approved analysis was undertaken at the University of North Carolina Children's Hospital and the Nemours Alfred I. duPont Children's Hospital covering the period from September 2000 through November 2006. Initially, all electronic and paper charts were examined in patients whose diagnosis was recorded as a CPT code of 742.3, 742.9, 741.0, or 742.4 as well as those with a radiographic report containing the words "Dandy" and "Walker." Patients with DWV were identified by the following radiographic characteristics: normal-sized posterior fossa, a posterior fossa cystic lesion that appeared to communicate with the fourth ventricle, and mild inferior vermian hypoplasia. The degree of vermian dysgenesis was graded using the following formula: length of vermis on the midline sagittal image/length of the cerebellar hemisphere on the midline sagittal image  $\times$  100). The vermian hypoplasia was graded as severe between 0 and 33%, moderate between 34 and 66%, and mild between 67 and 100%.

The posterior fossa size and volume was not rigidly measured because there are no standard criteria with regard to DWM or DWV, and therefore the senior author (A.M.R.) and study-dedicated neuroradiologist (V.J.) examined each film to determine whether they thought that the posterior fossa appeared to be consistent in size with that of patients without developmental anomalies.<sup>28</sup> Patients in whom the investigators observed mild vermian hypoplasia, subjective findings of a normal-sized posterior fossa, and a cystic lesion that openly communicated with the fourth ventricle were included in the study. The senior author and study neuroradiologist confirmed all radiographic diagnoses according to their experience and the general criteria set forth in diagnosing DWV.

Once the diagnosis of DWV was established, an exhaustive search of the electronic and paper hospital admissions, outpatient notes, and radiographic reports was undertaken. Anomalies including cardiac defects, craniofacial abnormalities, gastrointestinal abnormalities, genitourinary abnormalities, respiratory aberrations, and psychiatric issues as well as musculoskeletal dysmorphisms were assessed. The presence of a chromosomal defect or syndrome association was documented.

The senior author and the attending neuroradiologist examined the radiographs of all patients enrolled in the study for various radiographic abnormalities. Evidence of ventriculomegaly, vermian rotation as defined by the vermis extending superior to the posterior fossa cystic lesion, agenesis of the corpus callosum, occipital encephalocele, cortical atrophy, schizencephaly, syringomyelia, torcular inversion as defined by the confluence of sinuses being observed

above the lambdoid sutures, or polygyria, in addition to the overall characteristics that define DWV, were assessed.

The developmental status of the patients was evaluated based on an examination of the hospital and outpatient charts by the senior author or a dedicated pediatric neurosurgery nurse, or based on a physical examination by the senior author. The patients were of varying ages, so a standard outcome tool could not be used. Instead, modification of the Functional Independence Measure for Children and Disability Inventory instruments was undertaken. Specifically, the self-care domain, which includes eating, grooming, bathing, dressing, and toileting, was maintained. The mobility category of the Functional Independence Measure for Children was divided into no assistance, ambulatory with minimal assistance, moderate assistance, and maximal assistance (wheelchair, bedridden). The cognitive developmental domain was evaluated as functional comprehension and expression, social interaction, and schoolwork, as determined based on discussion with the family and review of reports from the child's school. The child was considered mildly delayed if he or she had significant problems in 1 category, moderately delayed if 2 systems were affected, and severe if delays were noted in all 3 domains.

The statistical analysis included general frequency analysis and two-tailed Pearson correlation. Direct-entry logistic regression analysis was attempted; however, results failed to converge. Regression diagnostics revealed a violation of assumption in a majority of cases. Therefore, results for the logistic regression analysis are not reported.

# Results

During the study period between September 2000 and November 2006, a total of 24 patients with DWV who were born between 1985 and 2005 were identified. Ten patients were boys and 14 were girls. The female-to-male ratio was 1.5:1. The mean gestational age was 35.6 weeks (range 23–41 weeks). Three patients were premature and were born at 23, 24, and 33 weeks. All other patients were born after 34 weeks of gestation. Three patients died. The mean follow-up period was 5.1 years (range 1 month–15 years). Nineteen patients (79.2%) were diagnosed based on findings of MR imaging studies, 4 based on CT scans, and 1 based on ultrasonography only.

#### Associated Congenital Anomalies

Extracranial and intracranial congenital anomalies were observed in 54% of patients (Fig. 1, Table 1). Cardiac abnormalities were seen in 41.7% of patients; defects included patent ductus arteriosus (7), ventricular septal defect (2), atrial septal defect (2), hypoplastic right heart (2), transposition of the great vessels (1), and mild pulmonary artery stenosis (1). Associated neurological conditions were observed in 33.3%; these included Grade III intraventricular hemorrhage, seizure disorder, cerebral palsy, hypotonia, progressive encephalopathy, autism, microcephaly, and cortical blindness. Gastrointestinal anomalies were observed in 20.8%, and included congenital diaphragmatic hernia, malrotation of the gut, omphalocele, tracheoesophageal fistula, and gastroesophageal reflux disease. Craniofacial anomalies were seen in 16.7%, and included cleft palate (2), cleft lip (1), hypertelorism (1), and Pierre Robin sequence with

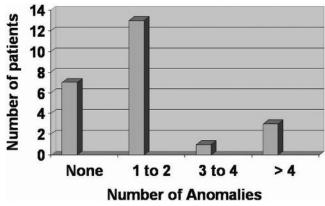


Fig. 1. Bar graph showing the incidence of anomalies associated with DWV. The x axis denotes the number of patients exhibiting 0, 1-2, 3-4, and > 4 associated anomalies, and the y axis denotes the number of patients.

mandibular and maxillary hypoplasia (1). Pulmonary, orthopedic, and genitourinary anomalies were observed in 12.5%. Chromosomal abnormalities were seen in 16.7%, and included partial trisomies of 9, 11, and 13 as well as mosaicism of chromosome 8. An associated syndrome was diagnosed in 12.5%.

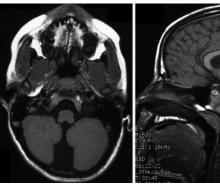
Ventriculomegaly, Hydrocephalus, and Radiographic

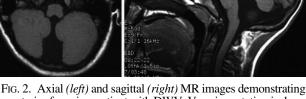
Vermian rotation was appreciated in 2 (8.3%) of 24 patients (Fig. 2 right). Other radiographic abnormalities including agenesis of the corpus callosum, which was observed in 5 (20.8%) of 24, and cortical atrophy in 3 (12.5%) of 24. Bilateral schizencephaly, a supratentorial arachnoid cyst, syringomyelia, polygyria, and torcular inversion were

TABLE 1 Incidence and description of associated abnormalities in 24 patients with DWV\*

System Involved	No. of Patients (%)	Type of Abnormality
cardiac	10 of 24 (41.7)	PDA, VSD, ASD, transposition, hypoplastic rt heart, pulmonary artery stenosis
neurological	8 of 24 (33.3)	Grade III IVH, seizures, CP, hypotonia, progressive encephalopathy, autism, microcephaly, cortical blindness
gastrointestinal	5 of 24 (20.8)	diaphragmatic hernia, malrotation of gut, omphalocele, TE fistula, GERD
craniofacial	4 of 24 (16.7)	cleft lip & palate, hypertelorism, man- dibular/maxillary hypoplasia
respiratory	3 of 24 (12.5)	restrictive lung disease, asthma, pul- monary hypertension
genitourinary	3 of 24 (12.5)	renal calculi, hydronephrosis, dysplas- tic kidney, end-stage renal disease
orthopedic	3 of 24 (12.5)	scoliosis, spasticity
chromosomal	4 of 24 (16.7)	trisomy 9, 11, 13, & mosaicism 8
associated	3 of 24 (12.5)	Senior-Loken, Smith-Lemli-Opitz,
syndrome		Pierre Robin sequence

<sup>\*</sup> ASD = atrial septal defect; CP = cerebral palsy; GERD = gastroesophageal reflux disease; IVH = intraventricular hemorrhage; PDA = patent ductus arteriosus; TE = tracheoesophageal; VSD = ventricular septal defect.





the posterior fossa in a patient with DWV. Vermian rotation is demonstrated in the sagittal orientation.

observed in 1 patient each, for an incidence of 4.2%. Occipital encephaloceles were not present in our cohort (Fig. 3). Ventricular enlargement was observed in 7 (29%) of 24 patients. Four of these 7 patients underwent placement of a CSF shunt system. The only variable that correlated with shunting was the presence of ventriculomegaly (p = 0.001). Of the 6 patients with isolated DWV, 3 required shunts.

# Developmental Status

Developmental outcome was assessed in only 21 patients because of early deaths in 2 children (< 2 months), and 1 who was lost to follow-up. Developmental delay was noted in 11 (52%) of these 21 patients. Six patients were severely affected, 2 demonstrated moderate developmental delay, and 3 were mildly affected. Five of 6 patients with isolated DWV had a normal developmental course. The sixth patient with isolated DWV exhibited developmental delay, and was slow to reach milestones. Her follow-up data were only available for 6 months before the family moved. Her ventricular enlargement did not significantly improve after shunt placement, and the opening pressure at surgery was low. Therefore, cortical loss may be a contributing factor. None of the 10 children who were developmentally normal had an accompanying neurological disorder. In contrast, in 7 of the 11 patients with developmental delay, the diagnosis was recorded as an associated neurological condition. When patients were grouped according to the absence or presence of developmental delay, the Pearson correlation determined that developmental outcome was solely affected by the presence of other neurological deficits (p = 0.001). The presence of an associated syndrome was also assessed (p = 0.081). When developmental delay was grouped into those who had none or mild delay versus those with moderate to severe delay, an associated neurological process remained significant (p = 0.026), and the presence of a syndrome was also important (p = 0.016).

### Discussion

Since its inception in the latter part of the 19th century by Sutton, with further refinements by Dandy in 1921, Walker in 1942, and Benda in 1954, various posterior fossa malformations have been characterized as the Dandy-Walker complex.<sup>5,7,11,12,29,34,35</sup> The complex is thought to represent a

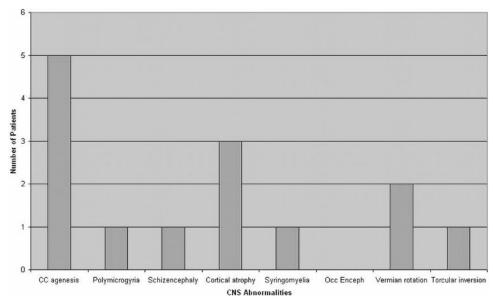


Fig. 3. Bar graph showing the associated radiographically confirmed CNS abnormalities observed in our population of patients with DWV. The y axis shows the number of patients in whom a particular abnormality was observed, and the x axis denotes the specific abnormality encountered. CC = corpus callosum; Occ Enceph = occipital encephalocele.

continuum of posterior fossa cystic abnormalities with varying degrees of vermian agenesis. The most well known, and the initial entity for which this complex attained its name, is the DWM.5,11,35 The DWM and its associated intra- and extracranial anomalies, incidence of hydrocephalus, and developmental outcomes are well established. 3,6,15-19,21,24,25,27,30,37 In 1976, Harwood-Nash and Fitz<sup>20</sup> described an addition to this spectrum, the DWV. This diagnosis is manifested by variable vermian hypoplasia, a normal-sized posterior fossa, and a cystic lesion that demonstrates open communication with the fourth ventricle. Specific size criteria to denote a normal-sized posterior fossa and vermian hypoplasia are not established in the literature. Rather, DWV is a diagnosis often made according to the expertise of individual neuroradiologists. There are only case reports of DWV in the literature. This is the first study in which an attempt has been made to expand our knowledge of this entity with respect to its correlation with associated congenital anomalies, radiographic abnormalities, hydrocephalus, and developmental outcomes.

The development of the posterior fossa structures is a well-orchestrated event. The cerebellum is formed by 2 distinct germinal matrices; 1 periventricular and 1 along the rhombic lip, which gives rise to the cerebellar hemispheres. 1,13,20,31 The cerebellar vermis develops as a thickening of the midline primordium of the rhomboencephalon during the 5th gestational week. By 16 weeks, the vermis fold and begins to cover the roof of the fourth ventricle. By 19 weeks of gestation the cranial/caudal length of the vermis is equal to that of the cerebellar hemisphere. The pathophysiological mechanism underlying the Dandy-Walker complex is not clearly elucidated. Initially, it was proposed that congenital obstruction of the foramina of Luschka and Magendie resulted in cystic dilatation of the fourth ventricle and the resulting malformed posterior fossa.<sup>20,34,35</sup> In later studies investigators have suggested that it is a manifestation of abnormal development of the rhomboencephalon, with incomplete formation of the vermis, or due to a defect within the tela choroidea, which leads to cystic dilation of the fourth ventricle. Given its comparable appearance to DWM, it is likely that DWV develops along the same embryological pathway.

#### Associated Congenital Anomalies

In the present cohort of patients with the DWV, the incidence of extracranial anomalies was 54%, compared with the 12-86% reported in patients with DWM.6,7,18,19,21,27,30 Multiple sonographic studies have demonstrated that associated anomalies occur in between 47 and 80% of fetuses with DWV and between 46 and 86% of those with DWM.<sup>8,15,16</sup> Cardiovascular abnormalities were the most frequently observed anomaly in our series. Only 1 other report exists in the literature correlating DWV with cardiac abnormalities, and it describes a 29-year-old woman with concomitant aortic coarctation, craniofacial anomalies, and DWV.2 Aberrations in the CNS, gastrointestinal, genitourinary, craniofacial, and musculoskeletal systems were found in this study. In our cohort, DWV was also associated with the Pierre Robin sequence, Smith-Lemli-Opitz syndrome, and Senior-Loken syndrome. In the literature, DWV has been associated with Menkes syndrome (kinky-hair disease), Coffin-Siris syndrome, and Ehlers-Danlos syndrome, as well as neurocutaneous melanosis. 4,22,23,26 Chromosomal abnormalities were observed in this study and included defects on chromosomes 9, 11, 13, and 8. Case reports have detailed observations of DWV and associated anomalies with deletions on chromosome 8; a long-arm deletion of chromosome 3 was observed in a patient with DWV and associated craniofacial anomalies; and partial trisomy 3 and monosomy 11 (a partial imbalance of chromosomes 6 and 11) have also been described. 9,14,33,38 The DWV may be associated with X-linked inheritance, as suggested by an ultrasound evaluation of a pedigree of 5 fetuses with isolated DWV, in whom only the males were affected.<sup>36</sup> Other studies have suggested a correlation with autosomal recessive inheritance, as was observed in a case of 2 siblings with concomitant diagnoses of DWV and spastic hereditary paraplegia.<sup>32</sup>

Ventriculomegaly, Hydrocephalus, and Radiographic Findings

The finding of ventriculomegaly and hydrocephalus in patients with the DWV has not been clearly ascertained. By comparison, a 55–96% rate of hydrocephalus has been observed in patients with DWM. 3.18,21,27,30 Based on prenatal ultrasonography studies, it has been estimated that ventriculomegaly will occur in 24–27% of patients with DWV. 15,16 In the current study, ventriculomegaly was observed in 29% of the patients, and approximately half required treatment with CSF diversion. The only factor that significantly predicted the need for a shunt was the presence of ventriculomegaly. Other radiographic features observed in our cohort, such as agenesis of the corpus callosum, gyral abnormalities, and cortical atrophy were comparable to those described in the literature for DWM. 3.18,21,27 Notably, no occipital encephaloceles were observed.

## Developmental Status

Poor developmental outcome in association with posterior fossa malformations is reported in 55-100% of patients. 17,25,37 It is estimated that moderate-to-severe developmental delay is observed in approximately one-third of patients with DWM, and of those 11–16% have a diagnosis of severe delay and require significant assistance in their daily functioning.<sup>3,6,18,21,27</sup> Developmental outcomes reported in patients with the DWV tend to be more positive. Based on a sonographic analysis, it was found that 7 of 13 patients in whom the DWV was diagnosed exhibited normal development.<sup>15</sup> However, the follow-up period in that study was only 6 weeks. In a retrospective sonographic analysis with longer follow-up ranging from 4 months to 4 years, investigators found that 9 of 11 surviving infants were developing normally, and that in 75% of those no associated extracranial sonographic anomaly was identified.<sup>16</sup> A radiographic study looking at the sensitivity of prenatal MR imaging found that 77% of infants were developmentally normal and 3 exhibited only mild motor delay, with a confirmed postnatal diagnosis of DWV.<sup>24</sup> Factors that appear to contribute to poor neurological outcomes in patients with DWM and DWV include the association with other CNS anomalies or neurological conditions. 6,37 In our study, a uniform developmental assessment was not feasible given the wide age range of the patients tested. A definitive correlation between developmental and intellectual outcome cannot be determined based on our results. However, our findings generally support the concept that isolated DWV is associated with a good outcome. The presence of other neurological anomalies or syndromes appears to increase the association with developmental delay.

## Conclusions

The DWV is a term that is frequently used in clinical practice to describe a milder form of the Dandy–Walker complex that refers to a constellation of findings including

a normal-sized posterior fossa, mild vermian hypoplasia, and a cystic lesion that communicates with the fourth ventricle. Rigid measurement criteria have not been established to ensure radiographic objectivity in making this diagnosis. However, it remains a frequent source of referrals for neurosurgeons.

Although our population of patients with DWV is small, it is the first study in which investigators have attempted to define the incidence of associated intra- and extracranial anomalies, radiographic findings, and developmental outcomes. We can conclude that DWV, like DWM, can be associated with other extracranial malformations, chromosomal abnormalities, and syndromes. Likewise, radiographic entities were observed with relative frequency in our study, calling into question the idea that DWV may occur along a developmental pathway similar to DWM. The incidence of associated hydrocephalus appears to be reduced in patients with DWV, but tends to be associated with ventriculomegaly, emphasizing the need for close follow-up of these patients to determine the possible need for CSF diversion. Developmentally, patients with isolated DWV appear to have a very good outcome when compared to the data quoted for those with DWM. The association with other neurological abnormalities appears to have a negative impact on the developmental outcome of these patients. In this study we found that in the DWV a spectrum of systemic anomalies can be observed. Radiographic abnormalities including ventriculomegaly and hydrocephalus do occur. When it is seen as an isolated entity, the overall developmental outcomes appear to be very good.

#### Disclaimer

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