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REVIEW ARTICLE

Dandy–Walker Syndrome (DWS): Clinical Features, Diagnosis, and Nursing Management

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ABSTRACT

Dandy–Walker Syndrome (DWS), also known as Dandy–Walker malformation (DWM), is a rare congenital brain malformation involving abnormal development of the cerebellum and fourth ventricle. The condition is commonly associated with hydrocephalus, developmental delay, motor dysfunction, and neurological impairments. The severity of symptoms varies widely depending on the degree of cerebellar involvement and associated congenital anomalies. Advances in prenatal imaging, neurosurgical interventions, and rehabilitative therapies have improved outcomes in affected children. Nursing management plays a critical role in early recognition of complications, neurological monitoring, developmental support, and caregiver education. This review article summarizes the etiology, pathophysiology, clinical manifestations, diagnostic approaches, treatment modalities, prognosis, and comprehensive nursing management of DWS.

KEYWORDS

• Dandy–Walker Syndrome • Cerebellar Malformation • Hydrocephalus • Prenatal Diagnosis • Neurological Assessment • Pediatric Nursing • VP Shunt • Developmental Support

INTRODUCTION

Dandy–Walker Syndrome (DWS) is a congenital malformation affecting the cerebellum and posterior fossa structures of the brain. The syndrome was first described by neurosurgeons Walter Dandy and Arthur Walker in the early twentieth century. DWS results from abnormal embryological development during the first trimester of pregnancy, particularly involving the cerebellar vermis and fourth ventricle.

The condition is characterized by hypoplasia or agenesis of the cerebellar vermis, cystic dilatation of the fourth ventricle, enlargement of the posterior fossa, and frequently hydrocephalus. The syndrome may occur as an isolated anomaly or as part of broader chromosomal or genetic disorders.

The estimated prevalence ranges from 1 in 10,000 to 1 in 35,000 live births, with a slightly higher prevalence among females.

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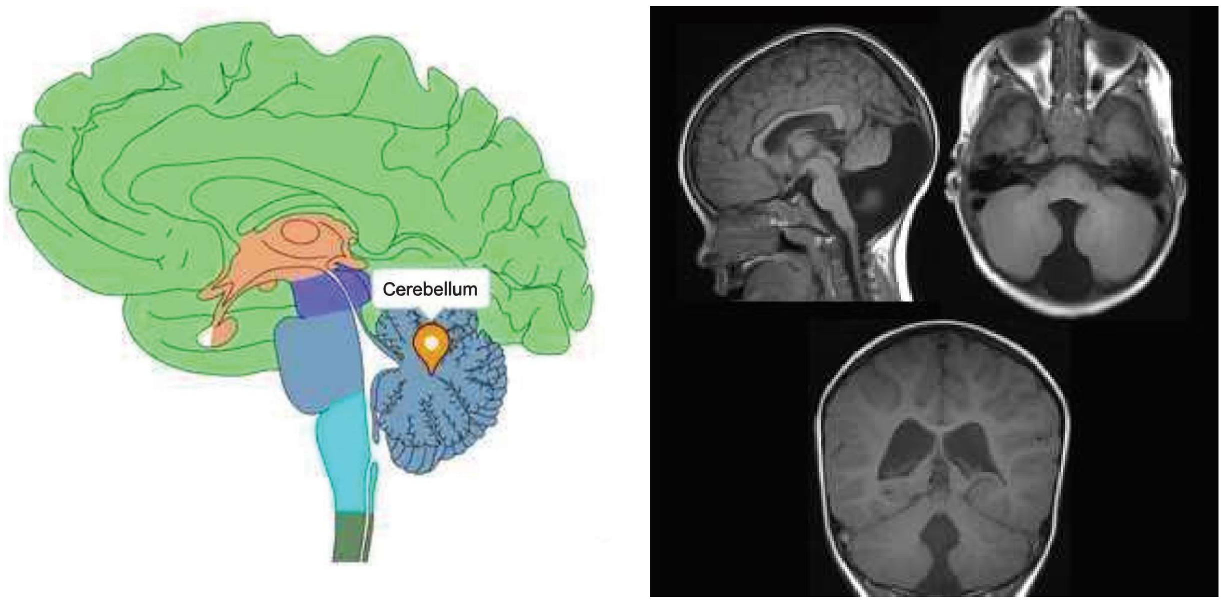


Figure 1: Normal Cerebellum vs Dandy–Walker Malformation, Comparison between a normal cerebellum and Dandy–Walker malformation showing hypoplasia or agenesis of the cerebellar vermis, enlarged fourth ventricle, and posterior fossa enlargement

ANATOMY AND PATHOPHYSIOLOGY

The cerebellum is a major structure located in the posterior part of the brain beneath the cerebral hemispheres and behind the brainstem. In normal anatomy and physiology, the cerebellum is composed of two hemispheres connected by the cerebellar vermis, which plays an essential role in maintaining balance, posture, coordination, muscle tone, and fine motor control. The fourth ventricle, situated between the brainstem and cerebellum, allows the normal circulation of cerebrospinal fluid (CSF) through the ventricular system and around the brain and spinal cord. The posterior fossa is the bony compartment that houses the cerebellum and brainstem. Proper development of these structures during embryonic life is necessary for normal neurological functioning, motor development, cognition, and coordination.

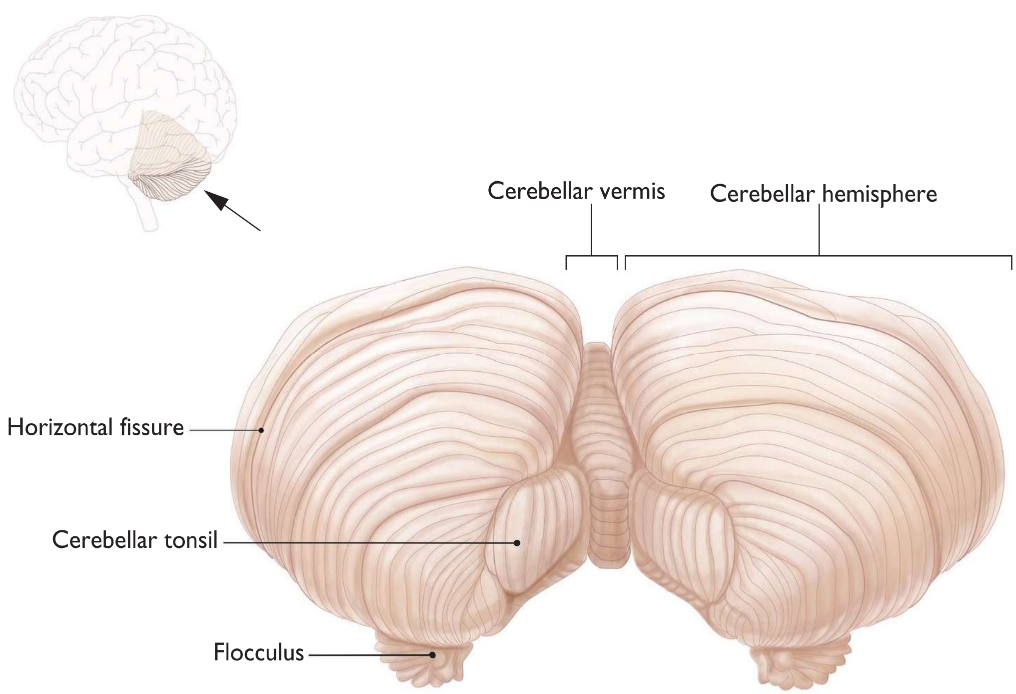


Figure 2: Anatomical Structures Affected in Dandy–Walker Syndrome

In Dandy–Walker Syndrome (DWS), abnormal embryological development of the cerebellum and surrounding posterior fossa structures occurs during early fetal life. The syndrome is characterized by partial or complete agenesis (absence) of the cerebellar vermis, cystic enlargement of the fourth ventricle, and enlargement of the posterior fossa. These abnormalities disrupt the normal flow and drainage of cerebrospinal fluid, frequently resulting in hydrocephalus and increased intracranial pressure. The impaired cerebellar development affects motor coordination, balance, posture, and muscle control, leading to delayed developmental milestones, ataxia, hypotonia, spasticity, and cognitive impairment in affected children. The severity of neurological dysfunction varies depending on the extent of cerebellar malformation and the presence of associated congenital anomalies.

The cerebellum is responsible for coordination, balance, posture, motor control, and aspects of cognition and behavior. In DWS, developmental disruption of the cerebellar vermis leads to impaired cerebellar connectivity and cerebrospinal fluid (CSF) circulation abnormalities.

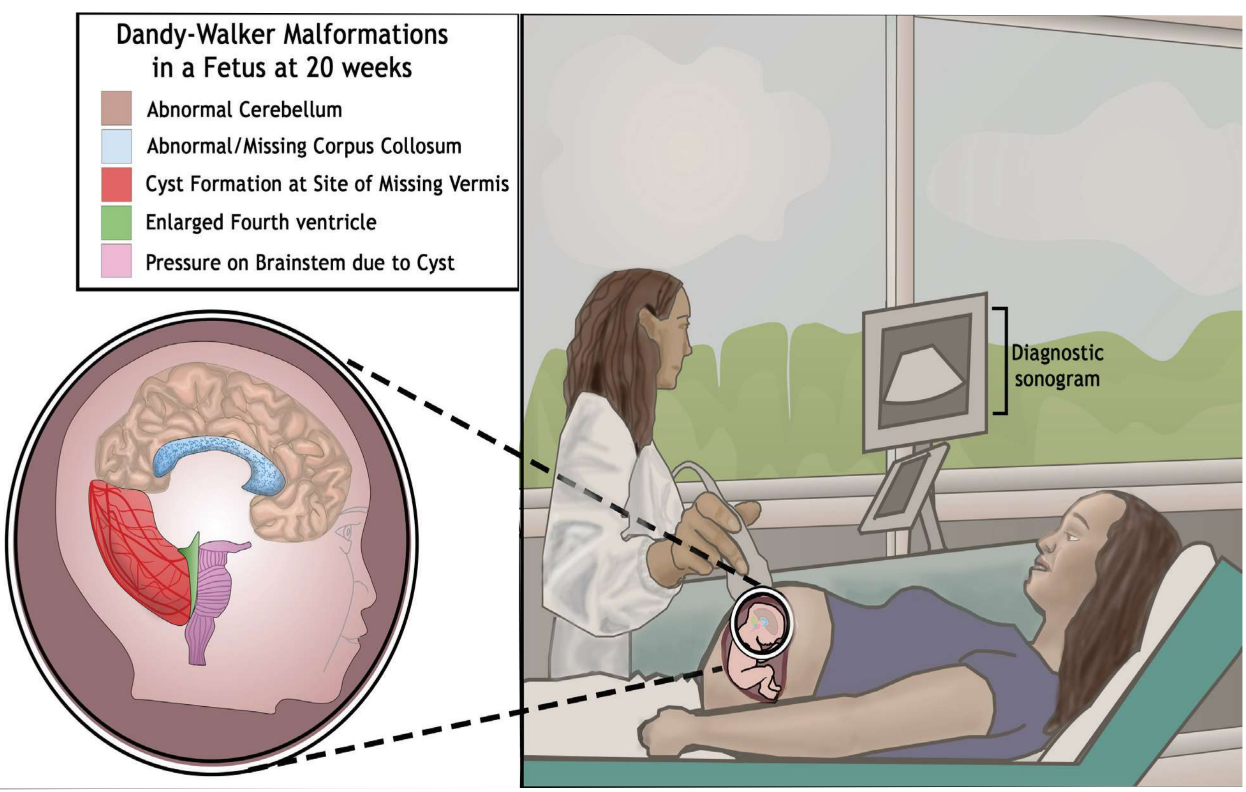
Major pathological features include:

1. Partial or complete agenesis of the cerebellar vermis
2. Enlargement of the fourth ventricle
3. Enlargement of the posterior fossa
4. Formation of a posterior fossa cyst
5. Hydrocephalus due to impaired CSF drainage

Hydrocephalus develops in approximately 70–90% of patients and significantly contributes to morbidity.

Etiology and Genetics

Dandy–Walker Syndrome (DWS) is considered a genetically heterogeneous congenital disorder, meaning that multiple genetic and chromosomal abnormalities may contribute to its development. Although the exact cause remains unknown in many patients, research has identified several genes and chromosomal defects associated with abnormal cerebellar and posterior fossa development during early embryogenesis. Among the most important genes implicated are the FOXC1, ZIC1, and ZIC4 genes, which play essential roles in brain and cerebellar formation. Mutations or deletions involving these genes can interfere with the normal development of the cerebellar vermis and cerebrospinal fluid pathways, leading to the characteristic malformations seen in DWS.



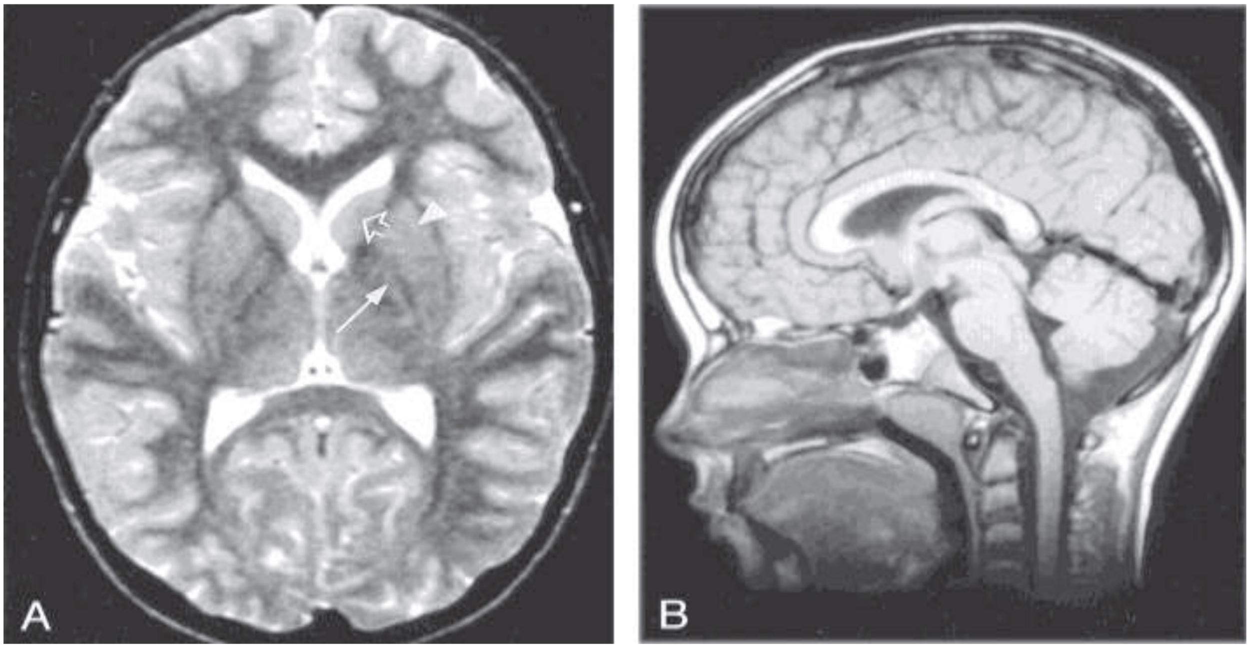


Figure 3: Prenatal Diagnosis of Dandy–Walker Syndrome. Prenatal ultrasonography and fetal MRI demonstrating posterior fossa abnormalities associated with Dandy–Walker Syndrome (***Source:*** embryo.asu.edu)

Chromosomal abnormalities are also strongly associated with the syndrome. DWS has been reported in individuals with trisomy 18, trisomy 13, trisomy 21, and trisomy 9, as well as in patients with chromosomal deletions or duplications affecting brain development genes. In some cases, DWS occurs as part of broader genetic syndromes such as Meckel syndrome, Warburg syndrome, and Ellis–van Creveld syndrome. Most cases are sporadic, occurring without a family history; however, familial occurrences suggest that both inherited and environmental factors may contribute to disease risk. First-degree relatives of affected individuals may have a slightly increased risk compared with the general population. Environmental influences such as maternal diabetes, prenatal infections, and exposure to teratogenic substances during pregnancy may further interact with genetic susceptibility and disrupt normal fetal brain development.

The precise etiology of DWS remains unclear; however, multiple genetic and environmental factors have been implicated.

Genetic Factors

Mutations associated with DWS include:

- FOXC1 gene mutations
- ZIC1 and ZIC4 deletions
- Chromosomal abnormalities such as trisomy 13, trisomy 18, trisomy 21, and trisomy 9

Environmental Factors

Potential prenatal risk factors include:

- Maternal diabetes mellitus
- Viral infections during pregnancy
- Exposure to teratogenic medications or toxins

DWS may also occur as part of syndromes such as:

- Meckel syndrome
- Warburg syndrome
- Ellis–van Creveld syndrome

Clinical features of Dandy–Walker Syndrome (DWS) arise primarily from maldevelopment of the cerebellum (especially the vermis), enlargement of the fourth ventricle, and progressive or congenital hydrocephalus. These structural abnormalities disrupt cerebrospinal fluid (CSF) circulation and impair cerebellar and brainstem function, leading to a wide spectrum of neurological and systemic manifestations.

Infant Presentation: Why These Symptoms Occur

Macrocephaly

Macrocephaly results from progressive accumulation of cerebrospinal fluid (CSF) due to obstruction of normal CSF flow at the level of the fourth ventricle. The enlarged ventricles increase intracranial volume, causing expansion of the skull, especially in infants whose cranial sutures are not yet fused.

Delayed Motor Milestones

Motor delay occurs due to cerebellar dysfunction, particularly involvement of the cerebellar vermis, which is responsible for:

- Postural control
- Trunk stability
- Coordination of early motor activities (sitting, crawling, walking)

Damage or underdevelopment leads to impaired timing and coordination of movements.

Hypotonia (Low Muscle Tone)

Hypotonia results from disrupted cerebellar modulation of motor pathways. The cerebellum normally facilitates muscle tone regulation via connections with the brainstem and spinal cord. When underdeveloped, muscle tone is reduced, leading to floppy infant presentation.

Spasticity

Spasticity occurs when there is involvement of upper motor neuron pathways or increased intracranial pressure affecting corticospinal tracts, leading to:

- Increased muscle stiffness
- Hyperreflexia
- Resistance to passive movement

Irritability and Vomiting

These symptoms are classic signs of increased intracranial pressure (ICP) caused by hydrocephalus. Elevated CSF pressure stimulates:

- Vomiting center in the brainstem
- Meningeal irritation
- Pain and discomfort due to brain stretching

Seizures

Seizures occur due to:

- Cortical irritation from increased intracranial pressure
- Associated cortical malformations (e.g., agenesis of corpus callosum)
- Secondary neuronal dysfunction due to disrupted brain organization

Neurological Manifestations: Mechanism-Based Explanation

Ataxia

Ataxia is caused by direct cerebellar vermis and hemispheric dysfunction, leading to:

- Loss of balance control
- Incoordination of voluntary movements
- Wide-based gait

Poor Coordination

Poor coordination results from impaired cerebellar integration of sensory and motor input, affecting:

- Fine motor control
- Timing of movements
- Smooth execution of voluntary actions

Nystagmus

Nystagmus arises due to cerebellar and brainstem involvement affecting vestibular control, leading to:

- Involuntary rhythmic eye movements
- Impaired gaze stabilization
- Visual tracking difficulty

Cranial Nerve Dysfunction

Compression or developmental abnormalities of the brainstem cranial nerve nuclei result in:

- Facial weakness
- Swallowing difficulty
- Eye movement abnormalities
- Hearing or vision impairment

Increased Intracranial Pressure (ICP)

ICP occurs due to obstruction of CSF flow from the fourth ventricle, leading to ventricular enlargement. This causes:

- Headache
- Vomiting
- Bulging fontanelle
- Lethargy
- Papilledema (in older children)

Associated Anomalies: Why They Occur

Agenesis of Corpus Callosum

This results from disrupted neuronal migration and midline brain development, often due to shared genetic mutations (e.g., FOXC1, ZIC genes). It leads to impaired interhemispheric communication.

Occipital Encephalocele

Occurs due to defective neural tube closure during early embryogenesis, allowing brain tissue to herniate through skull defects.

Cardiac Defects

Cardiac anomalies arise from common embryologic signaling pathways that affect both brain and heart development during early gestation.

Polydactyly / Syndactyly

These limb abnormalities occur due to genetic syndromes affecting embryonic patterning genes, often seen in syndromic forms of DWS.

Vision and Hearing Impairment

These result from:

- Cranial nerve involvement
- Brainstem dysfunction
- Associated cortical and structural anomalies

Late-Onset Presentation: Pathophysiology

Headaches

Headaches in older individuals result from chronic or compensated hydrocephalus, where CSF pressure intermittently increases.

Facial Palsy

Facial nerve dysfunction occurs due to brainstem compression or developmental abnormalities affecting cranial nerve VII pathways.

Behavioral Changes

Behavioral and cognitive changes are linked to:

- Cerebellar cognitive affective syndrome
- Frontal-cerebellar circuit disruption
- Chronic intracranial pressure effects on cortical function

Gait Abnormalities

Gait disturbances develop from progressive cerebellar dysfunction, leading to:

- Loss of balance
- Poor postural adjustment
- Unsteady or wide-based walking pattern

All clinical manifestations of Dandy–Walker Syndrome can be traced to three core mechanisms:

1. Cerebellar maldevelopment → coordination and motor deficits
2. CSF flow obstruction → hydrocephalus and increased intracranial pressure
3. Associated embryologic/genetic abnormalities → multisystem congenital defects

These combined disruptions explain the wide variability and severity of symptoms seen across patients.

Early and accurate diagnosis of Dandy–Walker Syndrome (DWS) is essential for timely medical intervention, surgical planning, developmental support, and family counseling. Diagnosis may occur during pregnancy through prenatal imaging or after birth when neurological symptoms and developmental delays become evident. A combination of radiological, neurological, developmental, and genetic evaluations is used to confirm the condition and identify associated anomalies.

Prenatal Diagnosis

Prenatal Ultrasonography

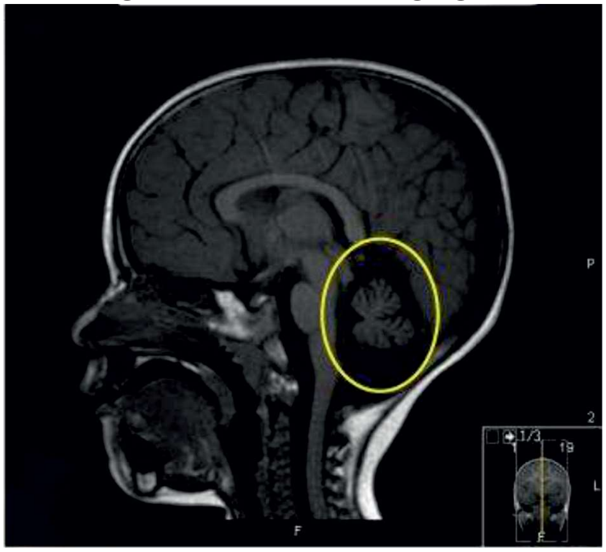
Prenatal ultrasonography is usually the first diagnostic tool used to detect DWS during the second trimester of pregnancy, commonly after 18–20 weeks of gestation. Ultrasound evaluation focuses on identifying abnormalities within the posterior fossa and cerebellum.

Detailed Findings on Prenatal Ultrasound

- Enlargement of the posterior fossa
- Cystic dilatation of the fourth ventricle
- Partial or complete absence of the cerebellar vermis
- Enlarged cisterna magna
- Ventriculomegaly or hydrocephalus
- Increased fetal head circumference
- Associated congenital anomalies such as cardiac defects or limb abnormalities

Ultrasound findings may vary depending on gestational age because the cerebellar vermis continues developing throughout pregnancy. Mild forms may be difficult to diagnose early.

Fetal Magnetic Resonance Imaging (MRI)



Fetal MRI provides more detailed visualization of fetal brain structures and is commonly performed when ultrasound findings are inconclusive or abnormal. MRI offers superior soft tissue contrast and better assessment of associated central nervous system abnormalities.

Detailed Findings on Fetal MRI

- Hypoplasia or agenesis of the cerebellar vermis
- Enlarged fourth ventricle communicating with a posterior fossa cyst
- Enlargement of the posterior fossa
- Elevation of the tentorium cerebelli
- Hydrocephalus or ventriculomegaly
- Corpus callosum abnormalities
- Brainstem abnormalities
- Cortical migration defects
- Associated neural tube defects

Fetal MRI is considered highly valuable for differentiating DWS from other posterior fossa abnormalities such as:

- Mega cisterna magna
- Arachnoid cyst
- Dandy–Walker variant

Computed Tomography (CT Scan)

CT scanning may be used in emergency settings or when MRI is unavailable. It helps assess hydrocephalus and bony abnormalities of the skull.

Detailed CT Findings

- Enlarged ventricular system
- Enlarged posterior fossa
- Cystic lesion in the posterior fossa
- Cerebellar hypoplasia
- Increased intracranial pressure signs
- Skull enlargement in infants with hydrocephalus

Although CT provides faster imaging, it is less detailed than MRI and involves radiation exposure.

Neurological Assessment

Clinical neurological examination is essential to identify functional impairments related to cerebellar and neurological dysfunction.

Detailed Neurological Findings

- Hypotonia or reduced muscle tone
- Spasticity in severe cases
- Poor coordination and balance (ataxia)
- Delayed motor milestones
- Abnormal reflexes
- Nystagmus or abnormal eye movements
- Cranial nerve abnormalities
- Seizure activity
- Signs of increased intracranial pressure:
  - Bulging fontanelle
  - Vomiting
  - Irritability
  - Lethargy
  - Enlarged head circumference

Neurological assessment also helps monitor disease progression and treatment response.

Developmental Screening

Developmental evaluation assesses the child's physical, cognitive, speech, behavioral, and social development.

Detailed Developmental Findings

- Delayed crawling and walking
- Speech and language delay
- Learning disabilities
- Intellectual disability ranging from mild to severe

- Poor fine motor coordination
- Behavioral and social interaction difficulties
- Delayed adaptive functioning

Standardized developmental screening tools and neuropsychological testing may be used for long-term follow-up.

Genetic Evaluation

Genetic counseling and chromosomal analysis are recommended, particularly in children with multiple congenital anomalies or family history of neurological disorders.

Genetic Testing Includes

- Karyotyping
- Chromosomal microarray analysis
- Fluorescence in situ hybridization (FISH)
- Gene mutation analysis for:
  - FOXC1
  - ZIC1
  - ZIC4

Detailed Genetic Findings

- Trisomy 18
- Trisomy 13
- Trisomy 21
- Chromosomal deletions or duplications
- Syndromic associations involving cerebellar malformations

Genetic evaluation helps:

- Determine recurrence risk
- Guide family planning
- Identify associated syndromes
- Improve prognostic counseling

Comprehensive diagnostic evaluation allows early multidisciplinary intervention, improves symptom management, and supports better developmental outcomes in children with Dandy–Walker Syndrome.

Top of Form

Bottom of Form

Medical and Surgical Management

Hydrocephalus Management

The primary surgical intervention is placement of a ventriculoperitoneal (VP) shunt to divert excess CSF and reduce intracranial pressure.

Seizure Management

- Anticonvulsant therapy
- Neurological monitoring

Rehabilitation

- Physical therapy
- Occupational therapy
- Speech therapy
- Developmental interventions

Multidisciplinary Care

Management requires collaboration among:

- Pediatricians
- Neurosurgeons
- Neurologists
- Rehabilitation specialists
- Nurses
- Psychologists
- Special educators

Nursing Management of Dandy–Walker Syndrome

Neurological Assessment

Nurses should:

- Monitor Glasgow Coma Scale (GCS)
- Assess muscle tone and reflexes
- Observe seizure activity
- Monitor signs of raised intracranial pressure

Signs of increased intracranial pressure include:

- Bulging fontanelle
- Vomiting
- Irritability
- Lethargy
- Enlarged head circumference

Monitoring Hydrocephalus and VP Shunt

Nursing responsibilities include:

- Monitoring shunt patency
- Assessing for infection
- Observing for CSF leakage
- Measuring head circumference regularly

Signs of shunt malfunction:

- Headache
- Vomiting
- Fever
- Decreased consciousness
- Seizures

Developmental Support

- Encourage age-appropriate stimulation
- Promote motor development
- Support communication skills
- Facilitate therapy participation

Nutritional Care

- Assess swallowing ability
- Monitor aspiration risk
- Ensure adequate hydration and nutrition

Family Education

Nurses should educate caregivers regarding:

- Disease process
- Warning signs of shunt malfunction
- Medication administration
- Developmental expectations
- Follow-up care

Psychosocial Support

Families often experience emotional stress and uncertainty. Nursing care should include:

- Emotional counseling
- Support groups
- Coping assistance
- Family-centered care

Complications

Major complications include:

- Hydrocephalus
- Recurrent seizures
- Developmental disability
- Respiratory complications
- VP shunt infection or obstruction

Complications related to hydrocephalus remain the leading cause of mortality.

Prognosis

The prognosis varies significantly depending on:

- Severity of cerebellar malformation
- Presence of hydrocephalus
- Associated genetic abnormalities
- Early intervention availability

Children with isolated DWS and timely intervention may achieve relatively normal cognitive and motor function. Severe associated anomalies often predict poorer developmental outcomes.

Prevention

Although DWS cannot always be prevented, risk reduction strategies include:

- Adequate prenatal care
- Maternal diabetes control
- Avoidance of teratogenic drugs
- Infection prevention during pregnancy
- Genetic counseling for high-risk families

CONCLUSION

Dandy–Walker Syndrome is a complex congenital neurological disorder involving abnormalities of the cerebellum and CSF pathways. Hydrocephalus, developmental delays, and neurological deficits are common features. Early diagnosis through prenatal and postnatal imaging, timely surgical management, and multidisciplinary rehabilitation are essential for improving outcomes. Nursing professionals play a pivotal role in neurological monitoring, developmental support, caregiver education, and psychosocial care. Comprehensive and family-centered management significantly enhances quality of life in affected children.

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