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Dandy - Walker syndrome: 3 cases report

Viroj Wiwanitkit*

Sithiporn Agthong**

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Dandy-Walker syndrome, a developmental anomaly of the nervous system, is a rare condition. Details about presentation, investigations, treatment associated programs and overall outcomes in 3 Thai patients were reported. The average age at diagnosis was 4 months, and the average maternal age was 24 years. Two cases with low birth weight and one case of preterm labor were observed. No cases with family histories of congenital anomalies were found. All cases were managed by ventriculoperitoneal (VP) shunt operation. No deaths occurred. The major post-operative complication in this study was due to shunt malfunction (2 cases, 66.6%). Other post-treatment complications were motor palsy (1 case), mental retardation (1 case) and convulsive disorder (1case). A case of child abuse was also detected in the study.

Key words: *Dandy-Walker syndrome, Anomaly.*

Reprint request : Wiwanitkit V, Department of Laboratory Medicine, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.

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* Department of Laboratory Medicine, Faculty of Medicine, Chulalongkorn University

** Department of Anatomy, Faculty of Medicine, Chulalongkorn University

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กลุ่มอาการ Dandy-Walker เป็นกลุ่มอาการที่มีภาวะรูปวิปริตของระบบประสาทเป็นกลุ่มอาการที่พบได้ไม่บ่อย ในการศึกษานี้ได้รายงานเกี่ยวกับประวัติอาการและอาการของผู้ป่วย การวินิจฉัย การรักษา ความผิดปกติที่พบร่วม ตลอดจนผลการรักษา ในผู้ป่วยชาวไทยจำนวน 3 ราย โดยอายุเฉลี่ยของผู้ป่วยขณะได้รับการวินิจฉัยประมาณ 4 เดือน อายุเฉลี่ยของมารดาประมาณ 24 ปี โดยเป็นรายที่มีน้ำหนักแรกคลอดต่ำกว่าปกติ 2 รายและคลอดก่อนกำหนด 1 ราย ทั้งนี้ไม่พบประวัติทางครอบครัวในผู้ป่วยทั้งหมด ผู้ป่วยทุกรายได้รับการรักษาโดยการผ่าตัดใส่ท่อเชื่อมระหว่างโพรงสมองและเยื่อบุช่องท้อง ไม่พบว่ามีผู้ป่วยรายใดที่เสียชีวิต ทั้งนี้ปัญหาที่พบมากที่สุดในการติดตามผลการรักษาได้แก่ ปัญหาการอุดตันของท่อเชื่อมที่ผ่าตัดไว้ (พบ 2 ราย คิดเป็นร้อยละ 66.6) สำหรับปัญหาอื่นๆที่พบได้แก่ ภาวะอัมพาตของกล้ามเนื้อ ภาวะปัญญาอ่อน และภาวะชัก โดยพบชนิดละ 1 ราย นอกจากนี้ยังพบการทารุณกรรมเด็กด้วยเป็นจำนวน 1 ราย

คำสำคัญ: *กลุ่มอาการ Dandy-Walker, ภาวะรูปวิปริต*

Dandy-Walker syndrome is a rare developmental anomaly of the nervous system. Abnormality of the rostral portion of the embryonic roof of the fourth ventrical with any degree of hypoplasia of the cerebellar vermis and medial aspect of the cerebellar hemisphere is the hallmark of this condition.[1] A hypothesis that the onset of the malformation occurs during genesis of the neural crest has been proposed.[2] High incidence of associated congenital disorders has been demonstrated.[2-5] Early diagnosis and treatment are necessary in the management of this condition.

This study reported 3 cases with Dandy Walker syndrome at King Chulalongkorn Memorial Hospital. Presentation, diagnosis, management and outcome of all cases were analyzed.

Case report

The complete records of 3 patients diagnosed with Dandy Walker syndrome at King Chulalongkorn Memorial Hospital were included in this report. Details about age at diagnosis, investigations, treatment, associated problems and overall outcomes were reviewed.

Medical history and prenatal history of each case were presented in Table 1. Average age at diagnosis was 4 months. It revealed the predominate incidence in female. All cases had developmental failure and hydrocephalus. No other associated anomaly was found. One case with history of Cesarian section due to cephalopelvic disproportion was found.

Table 1. Details of presentation of three cases of Dandy-Walker syndrome in this report.

	Patient 1	Patient 2	Patient 3
1. Medical history			
❑ Age at diagnosis	9 months	1 months	2 months
❑ Sex	male	female	female
❑ Chief complaint	delayed development	delayed development	delayed development
❑ Gross anomaly	hydrocephalus	hydrocephalus	hydrocephalus
❑ Family history of Congenital anomaly	no	no	no
2. prenatal history			
❑ Gestational	G1P1	G1P2	G1P1
❑ Maternal age	25 years	27 years	20 years
❑ Antenatal complications	No	history of valvular heart disease	No
❑ Pregnancy duration	Term	32 weeks	Term
❑ labor method	Normal labor	C/S	Normal labor
❑ Birth weight(gram)	3,950	1,610	2,300

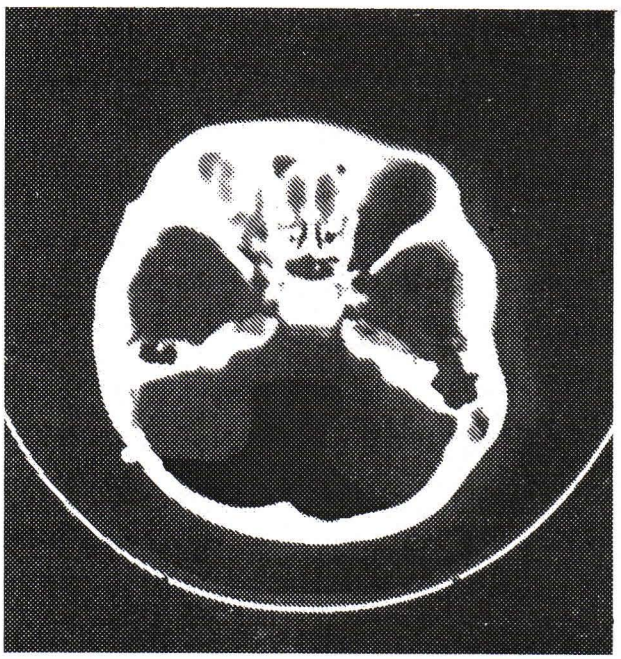

Figure 1. CT brain showed Dandy - Walker anamaly.

All cases were diagnosed by using CT scans of the brain (Figure 1). No prenatal diagnosis and chromosomal study was performed in all cases. Average maternal age was 24 years. Two cases with low birth weight and one case of preterm labor were noted. All cases were primigravid patients. No case with a family history of congenital anomaly was found.

The details of the clinical course of each case are presented in Table 2. All cases were managed by ventriculoperitoneal (VP) shunt operation. In our series, no deaths occurred. The major post-operative complication in this study was due to shunt malfunction (2 cases, 66.6 %). Numbers of shunt obstructions varied from 1 to 2 times. Other post-treatment complications were motor palsy (1 case), mental retardation (1 case) and convulsive disorder (1 case). A case of child abuse was also detected in our study.

Discussion

Dandy-Walker syndrome, a developmental anomaly first reported by Dandy and Blackfan in 1914,[6] is a rare neurological abnormality. Many sequeles, especially mental retardation and motor dysfunction can result from this condition. There was a slight, but insignificant female prevalence in our series (female: male = 2:1).

From our study, hydrocephalus was the common gross anomaly. This result is compatible with the study of Marinov et al,[7] which indicated the

Table 2. Details of clinical course of each case after treatment.

	Patient 1	Patient 2	Patient 3
1. Treatment	VP shunt	VP shunt	VP shunt
2. complications after treatment			
❑ Mortality	No	No	No
❑ Spastic motor palsy	No	No	Yes
❑ Intellectual retardation	No	No	Yes
❑ Seizure	No	Yes	Yes
❑ shunt malfunction	No	Yes	No
❑ child abuse	No	No	Yes

high incidence of hydrocephalus in this condition. Although about 50 % of cases associated with other anomalies were demonstrated in the previous study,[3] no case with other associated congenital anomalies was detected in this study. No familial association was detected.

From this study, the valid percentage of low birth weight (66.6 %) and Cesarian section (33.3 %) detected were similar to the previous study,[4] which showed 34 % and 44.7 % for low birth weight and Cesarian section, respectively. No multigravid or elderly pregnancy was detected in this study.

From the history review, the patients were brought to see the physician by their parents with the complaint of developmental retard edness and macrocephaly. Although no case could be prenatally or perinatally detected but all cases in this series were first diagnosed at an age below 1 year (average of 4 months). This aspect is rather early compared to the previous studies.[2 - 5]

All cases were managed by VP shunt operation and malfunctions of the shunt were detected in 66.6 % of total cases. Compared to the previous study, this type of shunt is not statistically different in malfunction rate to other types of shunt procedure.[8]

Although about 12.5 to 44.7 % of deaths occurred in the previous studies,[2 - 5] no mortality occurred in this study. Compared to the common sequeles, the incidences are lower (about 30 % for motor dysfunction and mental retardation) than in the previous studies.[2-5,9-10] Furthermore, in our study, a case of child abuse was also detected. This aspect should be considered in underprivileged groups of patients.

From literature review, there was no report about this syndrome in Thai patient before. The report is likely a useful basic information in management of this symptom. Data from the literature review of the Dandy- Walker syndrome in four previou non-Thai series[2-5] is also presented in Table 3.

Table 3. Data from literature review of case series of Dandy - Walker syndrome.

Series	Tai Y et al, 1980	Sawaya R et al, 1981	Hirsch JF et al, 1984	Pascual-Castroviejo I et al, 1991
Number of cases	21	23	40	38
Mortality (%)	48	26	12.5	44.7
Intellectual retardation(%)	72.7	71	40	58

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