Pielęgniarstwo Neurologiczne i Neurochirurgiczne
THE JOURNAL OF NEUROLOGICAL AND NEUROSURGICAL NURSING
eISSN 2299-0321 ISSN 2084-8021 www.jnnn.pl **Original**

DOI: 10.15225/PNN.2017.6.4.1

Health Problems of Children with the Dandy-Walker Syndrome

Problemy zdrowotne dzieci z zespołem Dandy-Walkera

Ewa Guz[1], **Renata Rogala**[2], **Dorota Nalepa**[3], **Elżbieta Bartoń**[4]

[1]Department of Neurological Nursing, Medical University in Lublin, Poland
[2]Medical Rescue Facility, Medical University in Lublin, Poland
[3]Department of Nephrology, Medical University in Lublin, Poland
[4]Department of Neurosurgery and Pediatric Neurosurgery, Medical University in Lublin, Poland

Abstract

Introduction. The Dandy-Walker syndrome is a complex congenital defect of the central nervous system, to which many health problems are related. In order to prepare oneself better to look after the child with this defect and their family, it is crucial to get to know the sickness unit itself, as well as the consequences of the related long-term neurological disorders.
Aim. Presenting health problems of children with Dandy-Walker syndrome.
Material and Methods. The study involved 30 ill children living in Poland. With the consent of parents, the medical records were examined. Additionally, the children were examined by pediatricians who assessed their condition and together with a survey, they documented it in the medical examination questionnaire.
Results. 7 children in the examined group are not able to function independently and they have an unpredictable prognosis of life expectancy. Only 3 children require periodic specialist care. The rest of the patients are constantly treated by various specialists both ambulatory and in hospitals. All children require regular, constant rehabilitation. It was examined in which group, girls or boys, there are more health problems. Both groups of 13 boys and 17 girls had the same number of diagnoses minimum 1 and maximum 5. The Mann-Whitney U Test did not show any relations statistically significant (p=0.83).
Conclusions. There is a large number of health problems indicating massive needs in care and hence, they require treatment by many specialists. Consequently, families of children with Dandy-Walker syndrome should be covered by active counselling on problems arising both from basic sickness unit and the coexistent ones. The family education should commence in the moment of diagnosis of the defect. **(JNNN 2017;6(4):138–142)**
Key Words: the Dandy-Walker syndrome, health problems, hydrocephalus, disability

Streszczenie

Wstęp. Zespół Dandy-Walkera jest złożoną wadą wrodzoną centralnego układu nerwowego, z którym związanych jest wiele problemów natury zdrowotnej. Aby dobrze przygotować się do opieki nad dzieckiem z tą wadą i jego rodziną, należy dokładnie poznać zarówno samą jednostkę chorobową jak też skutki długotrwałych zaburzeń neurologicznych z nią związanych.
Cel. Przedstawienie problemów zdrowotnych dzieci z zespołem Dandy-Walkera.
Materiał i metody. Badaniem objęto 30 chorych dzieci mieszkających na terenie Polski. Za zgodą rodziców przeanalizowano dokumentację medyczną. Dodatkowo dzieci zostały przebadane przez lekarzy pediatrów, którzy ocenili ich stan i wraz z pomiarami udokumentowali w kwestionariuszu badań lekarskich.
Wyniki. 7 dzieci z badanej grupy nie jest zdolnych do samodzielnego funkcjonowania z niepomyślnym rokowaniem, co do długości życia. Tylko 3 dzieci wymaga okresowej opieki specjalistycznej. Pozostałe są stale leczone przez różnych specjalistów zarówno ambulatoryjnie jak i w szpitalach. Wszystkie dzieci wymagają regularnej, stałej rehabilitacji. Sprawdzono, w której grupie, dziewczynek czy chłopców występuje więcej problemów zdrowotnych. W obu grupach 13 chłopców i 17 dziewczynek postawiono jednakową ilość diagnoz minimum 1 i maksimum 5. Test U Manna-Whitneya nie wykrył zależności istotnych statystycznie (p=0,83).

Wnioski. Ze względu na dużą liczbę problemów zdrowotnych wymagających leczenia przez wielu specjalistów, które wskazują na ogrom potrzeb w zakresie opieki, rodziny dzieci z zespołem Dandy-Walkera powinny być objęte czynnym poradnictwem w zakresie problemów wynikających zarówno z podstawowej jednostki chorobowej jak i tych współistniejących. Edukacja rodzin powinna rozpocząć się już w momencie rozpoznania wady. **(PNN 2017;6(4):138–142)**
Słowa kluczowe: zespół Dandy-Walkera, problemy zdrowotne, wodogłowie, niepełnosprawność

Introduction

The Dandy-Walker Syndrome is a very rare birth defect concerning hindbrain and including several characteristics which constitute to a fundamental recognition criterion. These traits include: fluid cyst on IV vault, agenesis or hypoplasia of cerebellar worm, overgrowth of holes draining cerebrospinal fluid with membrane septum (Magendi and Luschka) and higher localization of cerebellar hemispheres with hypoplasia in their structures [1–5]. There is also a syndrome variant in which all characteristics do not occur and it always accompanies other development or genetic defects. As the literature shows, 50 to 80% of cases are accompanied by hydrocephalus which requires implantation of pledge system [6–10]. Nowadays, in the era of development of diagnosis techniques, the Dandy-Walker Syndrome is recognized already in fetal life which gives possibility of preparing the family for welcoming a chronically ill child [6]. The cerebellum and the worm are responsible for regulation of muscle tension, balance and posture maintenance, control of neuromotics and as the latest studies indicate — for the higher cognitive functions [1,11,12]. Disorders resulting from the damages of those structures require constant, regular rehabilitation and early support in order to prevent from the formation of neurological defects. Treatment and rehabilitation need to be individualized and depend on the specific components of a particular case [7,11,13,14]. The goal of the study is to show the complexity of health problems of children with these defects and to indicate the areas in which the nursing personnel's support is indispensable.

Presenting health problems of children with Dandy-Walker syndrome.

Material and Methods

The study involved 30 children living in Poland. With a consent of parents, the medical documentation was analyzed. Additionally, children were examined by paediatricians who assessed their condition and together with measurements they documented this state in the questionnaire of medical examinations, a tool developed for the use of the Institute of Agricultural Medicine in Lublin. The tool was applied with a consent of authors.

Results

In the examined group, 22 children have full Dandy-Walker Syndrome with all the characteristics of this defect. The other 8 children were diagnosed with a syndrome variant. 13 children have epilepsy and 5 children suffer from cerebral palsy. Moreover, 2 children are treated due to the metabolic disorders and 3 have autism. 2 children have congenital disorder, asthma and Klippel-Feil syndrome. Such combination shows that one child can share both one and more chronic diseases which intensifies existing problems and hinders the treatment and rehabilitation of young patients. All examined patients have a disability certificate and chronic diseases are given as a cause (the Dandy-Walker syndrome does not have a statistical number). The examined children underwent different surgeries. 10 of them have had valve system implanted due to active hydrocephalus. Strabismus correction was made at in 6 children and other 4 are waiting for the procedure. Two children have had the meningocele operated and three have undergone heart surgeries. 3 children have had the tonsils removed. In individual cases, facial, oral and genital surgeries were performed as well as treatment of burn wounds or teeth removal under general anesthesia (2 cases) and closure of inguinal hernia. In total, 31 different surgeries have been performed. Due to the fact that all examined children have many chronic diseases, including birth defects, they need to be under constant care of specialized clinics. There are all clinics with the number of children for each of them listed in Table 1.

The majority of children, 93.3%, are under the care of neurological clinics. The other clinics with highest percentage of treated children, are the rehabilitation, neurosurgery, ophthalmic clinics. Again, it is worth emphasizing that each child can be treated in several clinics. The most frequent symptoms occurring in the study group are chronic headaches (40% of respondents). In the case of the birth defects of the nervous system, especially those related to the damages in cerebellum, the disorders in psychosocial development of young patients are often the case. The symptoms frequently aggravate already existing problems and the stress related to the staying in hospitals, painful treatment and social isolation, does not help. In the study group, 25 out of 30 children were diagnosed with abnormal psychosocial development. 18 children have speech abnormalities and some of them (6 persons) require intensive work with a speech therapist.

Table 1. List of specialist centres in which the examined children are treated

Type of specialist centre	N	%
Neurosurgical	15	50
Ophthalmology	14	46.6
Neurological	28	93.3
Logopedic	6	20
Psychological	9	30
Orthopedic	5	16.6
Rehabilitatio	16	53.3
Genetic	6	20
Laryngological	4	13.3
Gastrological	3	10
Allergic	5	16.6
Psychiatric	6	20
Endocrinological	7	23.3
Audiological	3	10
Cardiological	6	20
Nephrological	2	6.6
Urological	1	3.3

The analysis of the medical documentation showed that 7 children of the study group are not able to function independently, with a poor prognosis as to the life expectancy. Only 3 children require periodic specialist care. The rest of the children are treated by different specialists both ambulatory and in hospitals. The children were assigned with categories of care indicating what care they need. In the case of this study group, majority of children need medical (nurse) care as well as constant care of parents and other guardians because the system dysfunctions caused by particular diseases, do not allow independent existence.

Graph 1 shows the needs of examined children for service.

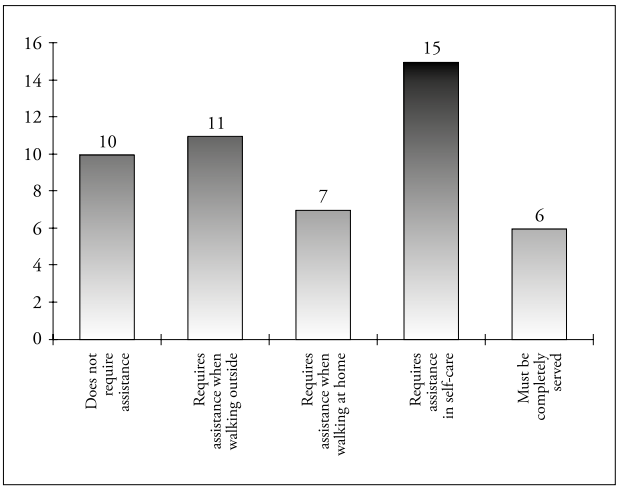

Figure. Needs of children for service

It was additionally examined, whether in a group of girls or boys there are more health problems. In both groups of 13 boys and 17 girls the same number of diagnoses of minimum 1 and maximum 5 were stated. The Mann-Whitney U test did not show any statistically significant statistically dependencies p=0.83. A similar situation took place when the children's age was considered. The dependencies are also not statistically significant here χ^2_{Yates} a=3.003, df=1, p=0.083.

Educating parents. As it was emphasized in introduction, it is crucial not to allow consolidation of neurological defects. It is possible only when parents work closely with the entire medical team and learn how to help a child in reaching the goal to function independently in society. Children with the Dandy-Walker syndrome tend to start to sit, walk and talk much later. In some cases, the ability to maintain balance and upright posture is reached at the age 3–5 years. This is the period in which cooperation with parents on the way to improve their children condition is indispensable. However, in order for this cooperation to be possible, parents have to acquire broad knowledge about the characteristics of the syndrome, results of occurring damages and types of rehabilitation supporting the psychomotor development of the child. The studies on the knowledge of patients' families about the Dandy-Walker syndrome were conducted in 2012 and published in 2014 [15]. The results indicated big gaps in knowledge particularly about the nature of the disease and the complications related to the occurrence of this defect. When we take into consideration this information, and the number of the health problems described, we can clearly see how much education in these areas is needed by the parents.

Comparison of Studies Results

As it was mentioned earlier, the Dandy-Walker syndrome is a very rare development defect CUN, which often coexists with other development and genetic defects. Subject literature describes cases of heart, skeletal defects and defects of the face. In the studied group there is one child with a heart defect not subject to a surgical correction. There is also one child with dysmorphosis and one with cleft lip and cleft palate. Over half of examined children underwent the diagnosis of metabolic disorders [16,17]. On the day of examination, one child had a confirmed diagnosis. In the article "Dandy-Walker malformation: analysis of 38 cases" I. Pascual-Castroviejo et al., described 25 defects and disorders coexisting with the Dandy-Walker syndrome in a group of 38 children [10]. Table 2 shows the defects that are shared by both groups.

Table 2. Comparison of defects coexisting in two examined groups with DWS

Name	Own studies group of 30 persons	Studies by I. Pascual-Castroviejo 38 persons
Macrocephaly	2 (6.6%)	31 (81.5%)
Hypertelorism	2 (6.6%)	14 (36.8%)
Strabismus	12 (40%)	14 (36.8%)
Psychomotorretardation	12 (40%)	8 (21%)
Intracranialhypertension	2 (6.6%)	6 (15.7%)
Palpebralptosis	1 (3.3%)	4 (10.5%)
Heartmalformation	1 (3.3%)	6 (15.7%)
Ophthalmicanomalies	1 (3.3%)	6 (15.7%)
Agenesis of corpuscallosum	3 (10%)	5 (13.1%)
Klippel-Feilsyndrome	1 (3.3%)	1 (2.6%)
Cleft lip and palate	1 (3.3%)	1 (2.6%)

The greater percentage of persons in own studies have psychomotor disorders and strabismus than in group of I. Pascual-Castroviejo. Yet, the macrocephaly was found in 81.5% respondents in group of I. Pascual-Castroviejo and only 6.6% in own study group. In the compared group 8 children were born as a result of the caesarean section while in own study group there were 14 such deliveries.

Conclusions

1. Due to a big number of health problems, which indicate the huge need for care, the families of children with the Dandy-Walker syndrome should be advised on issues arising both from the underlying disease unit and those coexisting.
2. Children with the Dandy-Walker syndrome require constant rehabilitation which means high financial costs. Hence, parents should receive all available information about institutions and foundations which support families with disabled children.

Implications for Nursing Practice

1. During the work with the families of children with damages of central nervous system, nurses should include the education of parents about the nature of the diseases, prevention of formation of neurological defects and ability to gain knowledge on the disease from different sources.
2. Education of families should commence when the defect is identified.

References

[1] Bolduc M.E., Limperopoulos C. Neurodevelopmental outcomes in children with cerebellar malformations: a systematic review. *Dev Med Child Neurol*. 2009;51(4): 256–267.
[2] Calabrò F., Arcuri T., Jinkins J.R. Blake's pouch cyst: an entity within the Dandy-Walker continuum. *Neuroradiology*. 2000;42(4):290–295.
[3] Lee A.Y., Jang S.H., Yeo S.S., Lee E., Cho Y.W., Son S.M. Changes in a cerebellar peduncle lesion in a patient with Dandy-Walker malformation: A diffusion tensor imaging study. *Neural Regen Res*. 2013;8(5):474–478.
[4] Manel L., Houneida Z.B., Habib A., Dejla B., Chekib K. A rare inborn error of metabolism associated with a Dandy-Walker malformation. *Acta Neurol Belg*. 2012; 112(4):425–426.
[5] Correa G.G., Amaral L.F., Vedolin L.M. Neuroimaging of Dandy-Walker malformation: new concepts. *Top Magn Reson Imaging*. 2011;22(6):303–312.
[6] Ulm B., Ulm M.R., Deutinger J., Bernaschek G. Dandy-Walker malformation diagnosed before 21 weeks of gestation: associated malformations and chromosomal abnormalities. *Ultrasound Obstet Gynecol*. 1997;10(3):167–170.
[7] Chang M.C., Russell S.A., Callen P.W., Filly R.A., Goldstein R.B. Sonographic detection of inferior vermian agenesis in Dandy-Walker malformations: prognostic implications. *Radiology*. 1994;193(3):765–770.
[8] Pilu G., Goldstein I., Reece E.A. et al. Sonography of fetal Dandy-Walker malformation: a reappraisal. *Ultrasound Obstet Gynecol*. 1992;2(3):151–157.
[9] Szymański W., Włodarczyk L. Zespół Dandy-Walkera: przegląd piśmiennictwa i opis przypadku. *Ultrasonografia Polska*. 1995;5(1):41–46.
[10] Pascual-Castroviejo I., Velez A., Pascual-Pascual S.I., Roche M.C., Villarejo F. Dandy-Walker malformation: analysis of 38 cases. *Childs Nerv Syst*. 1991;7(2):88–97.
[11] Krägeloh-Mann I. Lesions in early cerebellar development — do they matter? *Dev Med Child Neurol*. 2010;52(8): 694.

[12] Tavano A., Grasso R., Gagliardi C. et al. Disorders of cognitive and affective development in cerebellar malformations. *Brain*. 2007;130(Pt 10):2646–2660.
[13] Schmahmann J.D., Sherman J.C. The cerebellar cognitive affective syndrome. *Brain*. 1998;121(Pt 4):561–579.
[14] Kaczan T., Śmigiel R., Surowiec A. Dzieci z zespołem malformacji Dandye'go-Walkera. W: Kaczan T., Śmigiel R. (Red.), *Wczesna interwencja i wspomaganie rozwoju u dzieci z chorobami genetycznymi*. Wyd. Impuls, Kraków 2012; 135–144.
[15] Guz E., Turowski K. Zakres wiedzy rodzin pacjentów na temat zespołu Dandy-Walkera. *Piel. Zdr. Publ*. 2014; 4(2):127–134.
[16] Alafif M.M., Aljaid S.S., Al-Agha A.E. Central diabetes insipidus, central hypothyroidism, renal tubular acidosis and dandy-walker syndrome: new associations. *Ann Med Health Sci Res*. 2015;5(2):145–147.
[17] Dhupar V., Kumar P., Akkara F., Kumar A. Dandy Walker Syndrome with Tessier 7 Cleft-a Rare Case Report and a Surgical Note on the Use of Vermilion Flap and Lazy W-Plasty. *J Maxillofac Oral Surg*. 2012;11(3):368–370.

Corresponding Author:
Ewa Guz
Department of Neurological Nursing
Medical University in Lublin
ul. K. Jaczewskiego 8, 20-954 Lublin, Poland
e-mail: ewateresaguz@gmail.com

Conflict of Interest: None
Funding: None
Author Contributions: Ewa Guz[A–F, H], Renata Rogala[B, F], Dorota Nalepa[B, D, F], Elżbieta Bartoń[B, C, G]
(A — Concept and design of research, B — Collection and/or compilation of data, C — Analysis and interpretation of data, D — Statistical analysis, E — Writing an article, F — Search of the literature, G — Critical article analysis, H — Approval of the final version of the article)
Received: 03.09.2017
Accepted: 21.09.2017