·小儿神经外科·

Dandy-Walker综合征患儿腹腔分流术前后临床与影像学特征分析

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【摘要】 **目的** 总结Dandy-Walker综合征患儿腹腔分流术前后临床与影像学特征。**方法与结果** 选择2016年1月至2023年1月福建三博福能脑科医院收治的共6例Dandy-Walker综合征患儿(包括Dandy-Walker畸形1例、Dandy-Walker变型5例),主要表现为小脑蚓部缺损,小脑幕、窦汇区上移,第四脑室显著扩张,后颅窝囊性病变,幕上脑室扩张、积水(1例);或小脑蚓部发育不良,小脑半球受压,第四脑室扩张,后颅窝囊性病变,幕上脑室扩张、积水(5例)。分别行右侧侧脑室-腹腔分流术(2例)或后颅窝囊肿-腹腔分流术(4例),手术顺利,无手术相关并发症。随访2.31~7.00年,术后脑积水和后颅窝囊性畸形均逐渐减轻,3例预后良好,大脑半球和小脑发育良好,言语功能、运动功能、智力正常;3例预后一般,遗留轻度小脑平衡障碍(1例)或孤独症、幽闭恐惧症(1例),合并神经皮肤黑变病患儿遗留言语功能、运动功能、智力障碍(1例)。**结论** 表现为脑积水的Dandy-Walker综合征患儿经单分流术纠正脑积水后,大脑结构可获得良好发育,其中Dandy-Walker变型小脑蚓部良好发育,而Dandy-Walker畸形表现为小脑蚓部缺损时是否发育需更多病例观察。

【关键词】 丹迪-沃克综合征; 脑室腹膜分流术; 磁共振成像; 儿童

Clinical and imaging characteristics of children with Dandy-Walker syndrome before and after peritoneal shunt

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【**Abstract**】 **Objective** To summarize the clinical and imaging characteristics with Dandy-Walker syndrome (DWS) in children before and after peritoneal shunt. **Methods and Results** All 6 children with DWS admitted to Fujian Sanbo Funeng Brain Hospital from January 2016 to January 2023 were selected as the objects of observation, including one case of Dandy-Walker malformation (DWM) and 5 cases of Dandy-Walker variant (DWV). Before surgery, the main manifestations were cerebellar vermis defect, upward movement of the tentorial and sinusional area, severe dilatation of the fourth ventricle, posterior cranial fossa cystic lesions, supratentorial ventricle dilatation and hydrocephalus (one case); or cerebellar vermis dysplasia, cerebellar hemisphere compression, the fourth ventricle dilatation, posterior cranial fossa cystic lesions, supratentorial ventricle dilatation and hydrocephalus (5 cases). Right lateral ventriculoperitoneal shunt (2 cases) or posterior cranial fossa cyst-peritoneal shunt (4 cases) were performed, and the operations were successful without surgery-related complications. After 2.31–7.00 years of follow-up, 6 cases of hydrocephalus and posterior cranial fossa cystic malformation were gradually relieved, 3 cases of good prognosis, cerebral hemisphere and cerebellum development, speech, movement and intelligence normal; there were 3 children with general prognosis, including cerebellar disequilibrium symptoms (one case), autism, claustrophobia (one case), and speech, motor and intellectual disorders (one case) complicated with neurocutaneous melanosis. **Conclusions** The brain structure of the children with DWS presented with hydrocephalus can be well developed after the hydrocephalus is corrected by single shunt. Among them,

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the cerebellar vermis of the DWV is well developed, while the development of the DWM presented with cerebellar vermis defect needs to be observed in more cases.

【Key words】 Dandy-Walker syndrome; Ventriculoperitoneal shunt; Magnetic resonance imaging; Child

Conflicts of interest: none declared

Dandy-Walker综合征(DWS)是临床罕见的以小脑蚓部发育缺损或发育不良、第四脑室和后颅窝池扩张为特征的先天性中枢神经系统畸形,发病率为1/3.5万~1/2.5万[1],分为3种亚型,即Dandy-Walker畸形(DWM)、Dandy-Walker变型(DWV)和大枕大池[2-3]。外科治疗以腹腔分流术为首选,包括侧脑室分流术、囊肿分流术、侧脑室和囊肿双分流术[4]。由于Dandy-Walker综合征发病率较低,目前尚无大样本临床研究探究其病因、发病机制和长期预后。本研究拟对福建三博福能脑科医院近年诊断与治疗的6例儿童Dandy-Walker综合征患者的临床与影像学资料进行回顾总结,对比分析腹腔分流术前后脑形态改变,以期提高临床医师对疾病的诊断与治疗水平。

临床资料

一、纳入与排除标准

1. Dandy-Walker综合征不同亚型诊断标准 (1)Dandy-Walker畸形:小脑蚓部缺损或明显发育不良,第四脑室显著扩张,后颅窝池显著扩大且二者相通,小脑幕上抬[5]。(2)Dandy-Walker变型:小脑上蚓部正常、下蚓部部分缺失,第四脑室轻度扩张,后颅窝池扩张不明显,小脑幕位置正常[5]。(3)大枕大池:头部横断面MRI或CT检查显示枕大池直径>10 mm,小脑蚓部无异常,无脑室扩张和脑积水[6-7]。

2. 病例选择 (1)符合Dandy-Walker综合征各亚型诊断标准。(2)术前头部MRI检查显示合并脑积水。(3)为侧脑室-腹腔分流术或后颅窝囊肿-腹腔分流术后病例。(4)排除其他类型后颅窝囊性畸形、未合并脑积水且未行腹腔分流术以及临床资料不完善患儿。

二、一般资料

选择2016年1月至2023年1月在我院神经外科行腹腔分流术的Dandy-Walker综合征患儿6例,男性3例,女性3例;年龄22天至1岁7个月,平均0.65岁。临床分型包括Dandy-Walker畸形1例、Dandy-Walker变型5例;出生前后症状与体征表现为反复呕吐(2例),发作性肢体抽搐伴昏迷(1例)或发作性四肢强直(1例);胎儿期超声扫描脑发育异常(1例)或伴脑积水(1例),出生后头围进行性增大(3例)。术前影像学主要表现为小脑蚓部缺损,小脑幕、窦汇区上抬,第四脑室显著扩张,后颅窝囊性病变,幕上脑室扩张、积水(1例);小脑蚓部发育不良,小脑半球受压,第四脑室扩张,后颅窝囊性病变,幕上脑室扩张、积水(5例)。3例合并卵圆孔未闭、房间隔和室间隔缺损或右侧腹股沟斜疝和脐疝等畸形;1例伴神经皮肤黑变病。

三、治疗方法与结果

1. 腹腔分流术 根据术前矢状位T_2WI和磁共振相位对比电影(PC-MRI)所见,分别行侧脑室-腹腔分流术或后颅窝囊肿-腹腔分流术。(1)侧脑室-腹腔分流术:该术式主要用于矢状位T_2WI和PC-MRI显示中脑导水管通畅、幕上脑积水严重患儿。患儿仰卧位,气管插管全身麻醉,头偏向穿刺侧脑室对侧,于冠状缝前2.50 cm、中线旁开2.50 cm处做头皮直线或弧形小切口,将Codman可调压分流管(823114型,英特格拉生命科技制造公司)脑室端置入侧脑室额角,深度约5 cm。分流泵置于颞部,分流管腹腔端经胸腹皮下隧道引至上腹旁中线横切口,逐层切开皮肤、皮下组织、腹直肌鞘,分开腹直肌,于腹直肌后鞘和壁腹膜做一小切口,腹腔端分流管不剪裁全部置入腹腔。(2)后颅窝囊肿-腹腔分流术:适用于矢状位T_2WI和PC-MRI显示中脑导水管通畅、幕下脑积水严重患儿。患儿仰卧位,气管插管全身麻醉,头偏向拟置管侧对侧,于枕部旁正中做长约2.50 cm直切口,Codman可调压分流管脑室端置入囊腔内,深度3~4 cm;经皮下隧道将分流泵置于耳后,余步骤同侧脑室-腹腔分流术。

2. 随访与预后 (1)疗效评价标准:术后1个月行首次疗效评价,采用德国Siemens公司生产的3.0T MRI扫描仪(Magnetom skyra,24通道头部线圈)和Somatom Perspective 64排124层螺旋CT。MRI扫描参数包括T_2WI、T_1WI、FLAIR成像和可变反转角快速自旋回波(SPACE)序列。预后良好者,

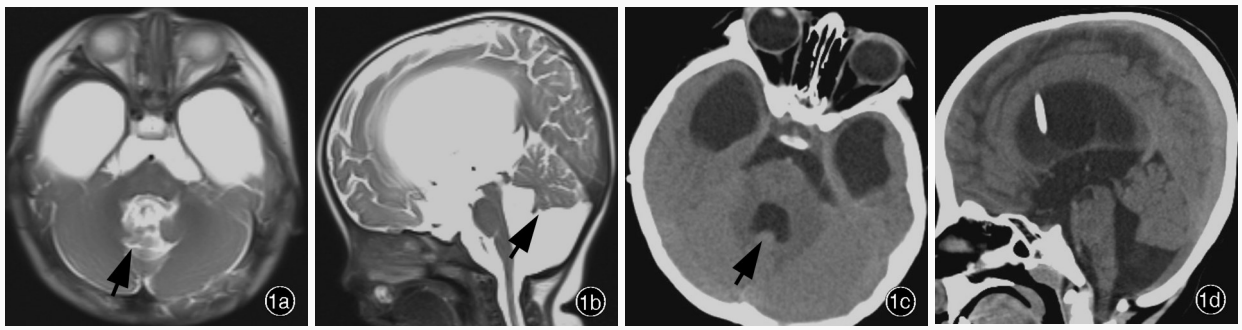

图1　男性患儿，9个月，因头围进行性增大3个月入院。临床诊断为Dandy-Walker变型，行右侧侧脑室-腹腔分流术。手术前后头部影像学检查所见　1a　术前横断面T_2WI显示小脑蚓部发育不良，后颅窝可见与第四脑室相通囊性信号影（箭头所示）　1b　术前矢状位T_2WI显示第四脑室扩张，小脑蚓部发育不良（箭头所示）　1c　术后1年5个月横断面CT显示小脑蚓部发育不良有所改善（箭头所示）　1d　术后1年5个月矢状位重建CT显示后颅窝囊性病变及幕上脑积水减轻，小脑蚓部及大脑半球发育改善

Figure 1　A 9-month-old male child was admitted to hospital with progressive enlargement of head circumference for 3 months. The clinical diagnosis was DWV, and right lateral ventriculoperitoneal shunt was performed. Pre- and post-operative head imaging findings Preoperative axial T_1WI showed dysplasia of cerebellar vermis, a cystic signal shadow communicating with the fourth ventricle was seen in the posterior cranial fossa (arrow indicates, Panel 1a). Preoperative sagittal T_2WI showed dilatation of the fourth ventricle and dysplasia of cerebellar vermis (arrow indicates, Panel 1b). Axial CT at one year and 5 months after surgery showed improvement in dysplasia of cerebellar vermis (arrow indicates, Panel 1c). Sagittal CT reconstruction at one year and 5 months after surgery showed posterior cranial fossa cystic lesions and supratentorial hydrocephalus were alleviated, and the development of cerebellar vermis and cerebral hemispheres were improved (Panel 1d).

影像学显示脑积水明显缓解、大脑皮质发育，临床检查无言语功能、运动功能、智力等神经功能严重障碍；预后一般，脑积水缓解、大脑皮质发育，但言语功能、运动功能、智力等神经功能明显障碍；治疗无效，脑积水无缓解，言语功能、运动功能、智力等神经功能明显障碍。(2)疗效与预后：本组患儿分别行后颅窝囊肿-腹腔分流术(4例)或右侧侧脑室-腹腔分流术(2例)，手术过程顺利，术后随访2.31～7.00年，平均(4.17±1.72)年；随访期间脑积水和后颅窝囊性畸形均逐渐减轻。①预后良好。3例患儿术后小脑蚓部发育不良明显改善、脑积水缓解(图1)，双侧大脑半球和小脑发育良好(图2)，且均无神经功能(言语功能、运动功能、智力)障碍。②预后一般。3例患儿术后脑积水缓解(图3)，其中1例随访5年孤独症、幽闭恐惧症表现持续存在，影像学检查不配合，运动功能发育尚可，仅上楼时平衡功能稍差，言语功能正常；2例术后随访2年6个月，癫痫发作得到有效控制，仅遗留言语功能、运动功能、智力障碍或轻度小脑平衡障碍、行走不稳。本组无治疗无效病例。

讨　论

1914年，Dandy和Blackfan首次报告1例13月龄患儿脑积水伴后颅窝囊肿和小脑蚓部发育不全，认为是炎症引起的Luschka-Magendie孔闭锁，命名为Luschka-Magendie孔闭锁症[8]；1942年，Taggart和Walker将其称之为先天性Luschka-Magendie孔闭锁，并发现窦汇区呈高位[9]；1954年，Benda报告6例Magendie孔闭锁病例，并指出Magendie孔闭锁症是一种发育异常，组织病理学表现为下髓帆被脑膜样囊取代、小脑分裂和部分畸形，并将其重新命名为Dandy-Walker综合征[10]。1989年，Barkovich等[11]经对Dandy-Walker综合征患儿临床与影像学资料的总结，认为Dandy-Walker畸形、Dandy-Walker变型和大枕大池等并非相互独立的疾病，而是后颅窝不同时期发育异常，因此提出“Dandy-Walker复合体(DWC)”这一概念以描述这种疾病连续体，并将后颅窝常见病变统一命名为Dandy-Walker复合体。Dandy-Walker复合体可以分为A型和B型两种亚型，A型包括Dandy-Walker畸形和Dandy-Walker变型，影像学可见小脑蚓部缺损或发育不良；B型包括Blake囊肿和大枕大池，小脑蚓部发育正常或接近正常，后颅窝囊状积液聚集区与第四脑室相通。

目前关于Dandy-Walker综合征的诊断存在两种观点：一种观点认为，不同病变类型的预后存在差异，应明确区分后颅窝不同囊性病变[12]，即鉴别诊断Dandy-Walker畸形、Dandy-Walker变型、Blake囊肿、大枕大池、后颅窝蛛网膜囊肿等各种后颅窝囊性病变。典型的Dandy-walker畸形影像学诊断标准包括[13]：(1)与第四脑室相通的后颅窝中线囊肿。

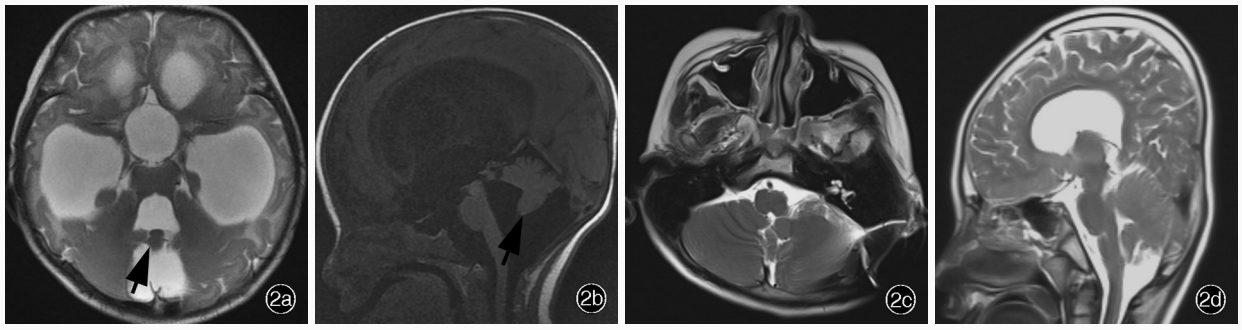

图2 男性患儿，2个月11天，因反复呕吐3天入院。临床诊断为Dandy-Walker变型，行右侧侧脑室-腹腔分流术。手术前后头部MRI检查所见 2a 术前横断面T_2WI显示小脑蚓部发育不良（箭头所示） 2b 术前矢状位T_1WI显示第四脑室轻度扩张，小脑蚓部发育不良（箭头所示） 2c 术后7年横断面T_2WI显示小脑发育良好 2d 术后7年矢状位T_2WI显示双侧小脑半球发育良好，小脑蚓部可见，后颅窝囊性病变基本消失

Figure 2 A 2-month-11-day-old male child was admitted to hospital due to repeated vomiting for 3 days. The clinical diagnosis was DWV, and right lateral ventriculoperitoneal shunt was performed. Pre- and post-operative head MRI findings Preoperative axial T_2WI showed dysplasia of cerebellar vermis (arrow indicates, Panel 2a). Preoperative sagittal T_1WI showed mild dilatation of the fourth ventricle and dysplasia of cerebellar vermis (arrow indicates, Panel 2b). Axial T_2WI at 7 years after surgery showed the cerebellum was well developed (Panel 2c). Sagittal T_2WI showed bilateral cerebellar hemispheres developed well, cerebellar vermis was visible, posterior cranial fossa cystic lesions basically disappeared at 7 years after surgery (Panel 2d).

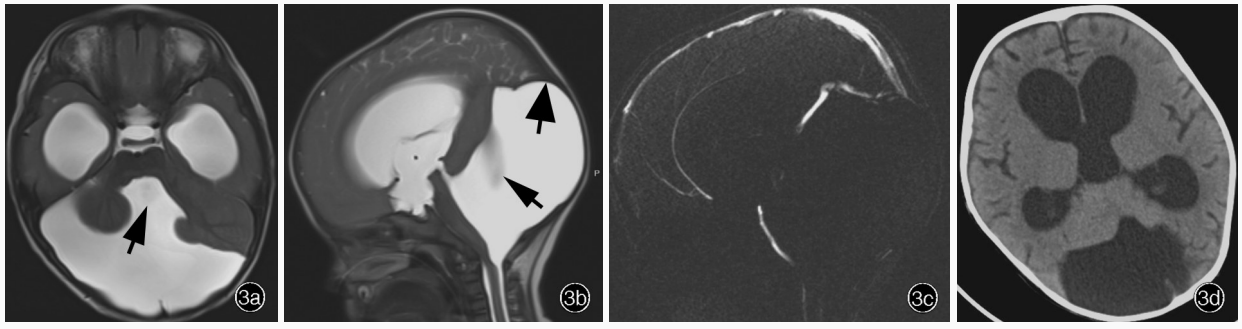

图3 女性患儿，1岁8个月，因反复呕吐5天、发作性肢体抽搐伴昏迷1天入院。临床诊断为Dandy-Walker畸形，行后颅窝囊肿-腹腔分流术。手术前后头部影像学检查所见 3a 术前横断面T_2WI显示小脑蚓部缺损（箭头所示），双侧小脑半球发育异常 3b 术前矢状位T_2WI显示中脑导水管通畅，小脑幕上抬（粗箭头所示），后颅窝可见与第四脑室相通囊性信号影（细箭头所示） 3c 术前PC-MRI未见中脑导水管和第四脑室脑脊液循环信号，桥前池脑脊液循环通畅 3d 术后2个月横断面CT显示脑积水缓解

Figure 3 A one-year-8-month-old female child was admitted to hospital due to repeated vomiting for 5 days and paroxyspasm with coma for one day. The clinical diagnosis was DWM, and posterior cranial fossa cyst-abdominal shunt was performed. Pre- and post-operative head imaging findings Preoperative axial T_2WI showed absence of cerebellar vermis (arrow indicates) and abnormal development of bilateral cerebellar hemispheres (Panel 3a). Preoperative sagittal T_2WI showed mesencephalic aqueduct was clear and upward tentorial movement of the cerebellum (thick arrow indicates), a cystic signal shadow communicating with the fourth ventricle was seen in the posterior cranial fossa (thin arrow indicates, Panel 3b). Preoperative PC-MRI showed no circulating signal of cerebrospinal fluid in the aqueduct and the fourth ventricle, and the circulation of cerebrospinal fluid in the anterior pontine cisterna was smooth (Panel 3c). Axial CT at 2 months after surgery showed relief of hydrocephalus (Panel 3d).

（2）不同程度的小脑下蚓部缺损。（3）小脑蚓部发育不良，残留蚓部向上、向前旋转移位。（4）第四脑室尖顶角缺失或变平。（5）后颅窝增大伴窦汇区上抬。（6）正常或发育不良的小脑半球前外侧移位。Dandy-Walker变型是一种Dandy-Walker畸形的不典型表现，第四脑室形成较好，扩张程度较轻，通常与蛛网膜周围腔相通；小脑蚓部发育不良较轻，后颅窝无明显增大。Blake囊肿由原始脉络膜外翻形成，病变与第四脑室相通，但与周围蛛网膜下腔并不相通[14]。大枕大池系小脑半球和小脑蚓部萎缩引起中线区脑脊液聚集，病变与第四脑室和蛛网膜下腔相通，故通常不会导致脑积水。后颅窝蛛网膜囊肿是位于蛛网膜层内的脑脊液聚集，不与蛛网膜下腔相通，病变对脑干和小脑存在占位压迫效应，小脑半球发育良好。值得注意的是，后颅窝蛛网膜囊肿需与孤立性第四脑室内蛛网膜囊肿相鉴别，后者临床罕见，小脑蚓部和第四脑室发育正常，囊肿体积大时第四脑室膨胀样扩张并向后压迫小脑，但正中孔无显著扩张[15]；前者病变与第四脑室、延髓脊髓蛛网膜下腔并不相通。后颅窝囊性病变改善有助于小脑发育，利于患儿运动功能、言语功能等神经功能发育，主要通过影像学检查相鉴别（表1）[16-18]。

表1 不同后颅窝发育异常亚型的神经影像学特征[16-18]

Table 1. Neuroimaging characteristics of different subtypes of posterior cranial fossa dysplasia[16-18]

分型	后颅窝	小脑蚓部	第四脑室尖顶角	第四脑室	脑积水	窦汇位置
Dandy-Walker畸形	扩大	缺损或发育不全	>45°	扩张	常见	上抬
Dandy-Walker变型	正常	不同程度发育不全	—	轻度扩张	可见	正常
Blake囊肿	正常	正常	<30°	扩张	常见	正常
大枕大池	正常	正常	正常	正常	无	正常
蛛网膜囊肿	正常	正常	正常	正常或缩小	可见	正常

—,not reported,未报道

另一种观点认为,Dandy-Walker畸形、Dandy-Walker变型、Blake囊肿、大枕大池属同一疾病不同亚型,可归于Dandy-Walker综合征或Dandy-Walker复合体,是后颅窝不同时期发育异常[17,19],Dandy-Walker复合体各亚型发生于妊娠第7~10周,尽管各亚型具有各自独特的解剖学特征,但在胚胎学中可被视为连续的统一体[20],各亚型具有相同胚胎起源。Ritscher等[21]报告一对具有相似颅面畸形的孪生姐妹,1例确诊为Dandy-Walker变型,另1例为大枕大池,进一步证实Dandy-Walker复合体各亚型具有相同胚胎起源。

目前,有关Dandy-Walker综合征的病因和发病机制尚不明确,多为散发,部分与单基因遗传病有关。有研究显示,环境、感染、代谢、母体因素等可导致胚胎发育过程中第四脑室正中孔、侧孔闭锁,小脑蚓部发育不良或者脑脊液动力学异常[22]。有80%~90%的Dandy-Walker综合征患儿可合并脑积水[13,23-24],Dandy-Walker综合征手术治疗主要依据是否需处理进展性脑积水或后颅窝占位导致的局部脑积水,故有学者将Dandy-Walker综合征视为一种特殊类型脑积水。根据中脑导水管、第四脑室、蛛网膜下腔通畅情况,Dandy-Walker综合征腹腔分流术可分为侧脑室-腹腔分流术、后颅窝囊肿-腹腔分流术、侧脑室及后颅窝囊肿-腹腔双分流术[10,25]。本研究基于影像学检查证实中脑导水管、幕上脑室与后颅窝囊性病变之间脑脊液循环无梗阻,单分流术即可控制病情,并根据幕上、幕下脑积水严重程度,选择脑积水严重处放置分流管。本组6例患儿均采用单分流术,术后脑积水明显缓解,有1例患儿入院时影像学显示小脑蚓部发育不良,术后7年影像学显示小脑蚓部发育正常,提示Dandy-Walker变型患儿的小脑蚓部发育不良可能为后颅窝局部囊状积液压迫所致,局部积液得到有效控制后小脑蚓部即可逐渐恢复至正常发育过程,但是否所有Dandy-Walker变型均可恢复至正常,尚待大样本长期随访研究证实。仅有的1例Dandy-Walker畸形患儿治疗有效,但因表现为孤独症、幽闭恐惧症而无法配合远期影像学随访,未能明确影像学表现为小脑蚓部缺损时小脑蚓部是否发育,今后尚待更多病例观察。

神经皮肤黑变病患儿通常可合并多种中枢神经系统畸形,尤以Dandy-Walker畸形常见,8%~10%的神经皮肤黑变病患儿可出现Dandy-Walker畸形[26]。本组有1例患儿合并神经皮肤黑变病,经后颅窝囊肿-腹腔分流术治疗,癫痫发作得到有效控制,但言语功能、运动功能、智力等神经功能障碍仍然存在。有研究显示,合并神经皮肤黑变病的Dandy-Walker畸形患儿预后较差[26-27],本研究该例患儿术后神经功能障碍未得到有效改善可能即与合并神经皮肤黑变病有关;此外,囊肿体积和合并其他中枢神经系统异常是Dandy-Walker畸形患儿预后不良的重要预测因素[4]。Sun等[1]的随访研究共纳入19 506例妊娠期女性,76例经超声确诊胎儿患有Dandy-Walker综合征,其中19例(25%)为孤立性Dandy-Walker综合征(孤立组)、57例(75%)合并其他部位畸形(合并组),孤立组9例终止妊娠、7例继续妊娠、3例失访,所有胎儿出生后均未见明显异常;合并组44例终止妊娠、12例继续妊娠、1例失访,继续妊娠者部分胎儿出生后表现有不同程度异常,包括神经发育迟缓(1例)、出生后1周死亡且伴多器官发育异常(1例)、严重感音神经性聋(1例)、失访(1例),但大多数(8例)患儿发育正常,提示合并其他部位畸形的Dandy-Walker综合征患儿可能预后更差。

综上所述,表现为脑积水的Dandy-Walker综合征患儿通过单分流术纠正脑积水后可使小脑蚓部

及大脑结构获得良好发育,今后将扩大样本量并对患儿进行长期随访,以进一步明确腹腔分流术在Dandy-Walker综合征治疗中的价值。

利益冲突 无

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