

Diagnosing Dandy-Walker Complex by Computed Tomography: Experience in Uganda and Recommendations for Hospitals in Resource-Limited Settings

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Clinicians in the developing world – particularly those treating infants and patients with congenital neurological diseases – should be equipped with the knowledge to diagnose Dandy-Walker complex (DWC) without relying on solely magnetic resonance imaging (MRI). In this paper, we will outline our recommendations for accurate diagnosis of DWC using computed tomography (CT) imaging on a sample of East African patients treated at our hospital in Uganda. Finally, we present a closer look at the social obstacles faced by this unique disease, and offer a framework for approaching these challenges.

What is the Dandy-Walker Complex?

“Dandy-Walker malformation” was first used in 1954 to describe the combination of a cystic dilation of the fourth ventricle and a hypoplastic cerebellar vermis (Benda, 1954). Since that time, the eponym has undergone modifications. Citing inherent challenges in the diagnosis of individual disease entities and inconsistencies within the literature base, Barkovich and colleagues introduced the term Dandy-Walker complex (DWC) in 1989 to represent a continuum of developmental anomalies of the posterior fossa (Barkovich, Kjos, Norman, & Edwards, 1989). The members of DWC each have varying degrees of fourth ventricular cystic dilation, vermian dysgenesis, and posterior fossa enlargement, as described in the sections that follow. In increasing severity, the DWC includes mega cisterna magna (MCM), Blake's pouch cyst (BPC), Dandy-Walker variant (DWV), and Dandy-Walker malformation (DWM). DWC is a relatively common congenital malformation occurring in at least 1 in 5,000 liveborn infants in the United States (Parisi & Dobyns, 2003). Though not fully elucidated, DWC likely results from a defect in the embryologic development of loose connective tissue of the pia mater within the fourth ventricle, creating a dorsal outpouching and variable vermian hypoplasia. The result is often an obstructive form of hydrocephalus secondary to inadequate fourth ventricle cerebrospinal fluid (CSF) drainage.

How Does DWC Present?

In the developing world, DWC usually comes to medical attention following a protracted period of hydrocephalus that eventually leads to overt macrocephaly. The majority of our patients in Uganda had a head circumference exceeding the 95th percentile at roughly 3.5 months of life. Many patients

were also present with signs and symptoms of chronic increased intracranial pressure, which include irritability, vomiting, bulging fontanelle, gaze palsy, and nystagmus. Though structural involvement of the cerebellum, in part, defines DWC, cerebellar dysfunction in infancy is uncommon (Kumar & Burton, 2008).

Imaging Techniques and Diagnosis in Underdeveloped Countries

Autopsy analysis aside, imaging is requisite for diagnosis of DWC. Western medicine has the benefit of routine prenatal sonographic evaluation of fetal well-being, and therefore, the majority of patients receive neurosurgical consultation immediately after birth. Patients receive MRI evaluation to confirm the prenatal diagnosis of DWC and to identify associated central nervous system (CNS) anomalies, which are present in up to 71% of DWM patients (Has et al., 2004). The superiority of MRI over computed tomography (CT) scans in evaluating the posterior fossa structures means that few, if any, DWC patients in the U.S. will receive a CT scan. MRI has the advantage of defining in detail the precise nature of CNS abnormalities.

Such luxuries are distant considerations for many patients in underdeveloped countries. The next best option for these patients is a CT scan. Though commonplace even among rural hospitals in the United States, a CT scanner can nonetheless be a rare commodity outside the developed world. Take Uganda as an example: as of early 2011, there were only 3 CT scanners outside Uganda's capital city. Poor road networks, rising fuel prices, and limited finances collectively create a challenging dilemma for patients who live far from a CT scanner.

In the case of MRI, many third world countries like Uganda don't even have a MRI device. In countries that *do* have MRI devices, MRI use is often cost-prohibitive. Whereas a single CT scan can cost as little as \$60 USD, the cost of MRI, on average, exceeds \$500 USD; such an expense is arguably excessive for impoverished populations. While modern medicine may encourage the use of MRI for accurate diagnosis of DWC, there needs to be an alternative for the developing world; a CT scan is a more reasonable, cost-efficient option. With adequate support, most patients can receive a CT scan. Based on our experiences working with patients at the Cure Children's Hospital of Uganda (CCHU), even patients from the most desperate of socioeconomic and political environments have been able to obtain CT scans.

Our Experience and Recommendations

In 2004, the United States Agency for International Development (USAID) donated a CT scanner to Cure Children's Hospital of Uganda (CCHU). This asset significantly improved the diagnostic capabilities of a hospital treating diseases that demand an advanced imaging modality. Because of this acquisition, clinicians at the hospital are now able to treat dozens of diseases that previously went undiagnosed.

Dandy-Walker complex is one such disease. Between 2004 and 2010, 45 patients were diagnosed with and treated for DWC at CCHU. These patients were selected from a population of 143, identified as having cyst-like posterior fossa pathology or hindbrain dysplasia. It was from this larger population that the diagnostic criteria, outlined below, were applied to extract those with DWC. The following are the three disease entities along the DWC spectrum that we feel can accurately be diagnosed using CT imaging. Our recommendations for accurate diagnosis combine features of accepted criteria outlined elsewhere (Sasaki-Adams et al., 2008; Strand, Barnes, Poussaint, Estroff, & Burrows, 1993).

Dandy-Walker malformation (DWM)

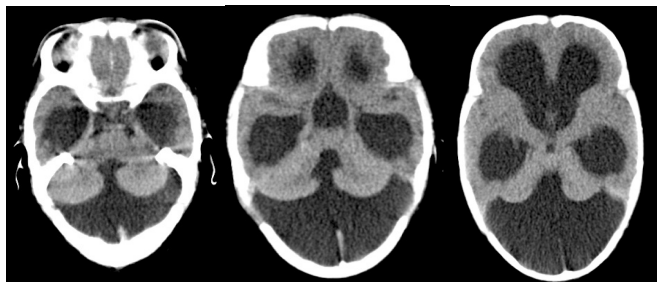
1. Complete absence of the cerebellar vermis, or severe unequivocal hypoplasia of the inferior vermis
2. A posterior fossa CSF collection in direct communication with the fourth ventricle
3. An unequivocally enlarged posterior fossa

Dandy-Walker variant (DWV)

1. Inferior vermis hypoplasia
2. Posterior fossa CSF collection in direct communication with the fourth ventricle
3. No obvious enlargement of the posterior fossa

Mega cisterna magna (MCM)

1. Enlarged cisterna magna
2. Large posterior fossa volume
3. Formed cerebellar vermis



Dandy-Walker Malformation. Axial CT images reveal significant posterior fossa enlargement, absence of the cerebellar vermis and dilation of the third, fourth, and frontal and temporal horns of the lateral ventricles.

We feel that the above characteristics can all be accurately identified or refuted by axial CT imaging, and do not require MRI. The features above can be recognized and documented by a physician with a basic grasp of neuroimaging. It goes without saying, however, that any clinician responsible for reading such images should have a firm understanding of neuroanatomy. These points are not meant to suggest that the diagnosis of and distinction between individual DWC entities are simple tasks. While trained neurosurgeons at CCHU are the ones who read CT scans, a general physician can also obtain such scans and identify features that suggest a diagnosis

of DWC. The patient, and the CT images, should then be sent to a physician who has the ability to treat the pathology. Before an intervention can be performed on such patients, an evaluation of clinical and radiographic data must always be performed by the physician who intends to intervene surgically. From a general physician's perspective, absolute diagnostic certainty is not necessary. A well-informed suspicion of the presence of DWC based upon CT findings would suffice.

In our experience, each of the 45 patients were present with macrocephaly with or without signs of increased intracranial pressure. Following diagnostic imaging with CT, patients were classified as DWM, DWV, or MCM, and they were surgically managed for resolution of hydrocephalus. Endoscopic third ventriculostomy and/or ventriculoperitoneal shunting achieved resolution of signs and symptoms of progressive hydrocephalus in the vast majority of patients.

Special Considerations for DWC Patients

Proper management of these unique patients requires commitment from a large and diverse group of healthcare providers including pediatricians, radiographic technicians, nurses, anesthetists, neurosurgeons, and laboratory personnel. In an environment where financial and human resources are limited, often a single individual may occupy many of these roles. Furthermore, while the medical care these patients receive while hospitalized is of utmost importance, the social environment surrounding the patient and its implications for a successful recovery cannot be overstated.

Hydrocephalus, whether associated with DWC or another cause, carries a powerful stigma in many cultures; it is a stigma that poses great barriers to patient recovery and societal assimilation. In Uganda, for example, a mother might perceive a child with macrocephaly as an 'alien,' 'demon,' or the 'devil' himself. Some fathers believe that such children are the consequence of infidelity on the part of the mother. In fact, the fathers of the majority of macrocephalic children treated at CCHU have abandoned their spouse. Communities will often reject the mother and child for fear that either one carries a negative contagion that could disrupt society. Unfortunately, some mothers will even kill their children in an effort to escape the burden or rid her community of a presumed evil being.

It is for these reasons that the hospital maintains an active and well-supported social outreach team charged with educating patient caregivers about their child's disease, and offering spiritual, social, vocational, and occasionally, financial support. It became apparent early in the establishment of CCHU that surgical outcomes correspond with the degree of social support surrounding a patient and his or her caregiver(s). During the hospital admission process, trained social workers evaluate each caretaker's social status, including his or her home environment, level of education, occupation, spiritual needs, and community support. During this evaluation, caretakers often paint a picture of the hardships suffered during the life of their child. A plan is then made for each patient, and social workers, religious staff, and medical personnel work in concert to address these challenges and develop solutions.

At CCHU, a large effort is made to educate patient caretakers about the etiology of their child's condition. Through words, pictures, and videos, the medical staff offers a basic ex-

planation for the biological cause of their child's disease. This knowledge is meant both to eliminate the caretaker's fear that their child is a form of punishment and to empower him or her with knowledge to educate community members at home.

Religious leaders employed by the hospital are available to counsel caregivers and their children throughout their hospital stay. Families are free to interact as much or as little as they choose with the religious staff. Spiritual counseling is available to guests on an individual basis, and prayer and song sessions are regularly held for community worship.

CCHU strives to accommodate patients from every geographic region and tribe of Uganda and beyond. With over 80 different tribes and scores of distinct languages and dialects, CCHU employs staff that speak dozens of languages to ensure that patient caretakers can tell their child's story in their native tongue. The hospital leadership believes that of all the obstacles faced by these patients, a language barrier is one that can be eliminated from the outset. Finally, CCHU empowers patient caretakers, and women in particular, by equipping them with 'income-generating' activities. A section of the hospital is charged with training women to make beaded necklaces from scrap paper, and selling their work in local markets. Such activities are designed to provide patients with a small source of income and to reduce their dependence on potentially unreliable partners or family members.

This multifaceted approach to caregiving makes CCHU unique among Ugandan hospitals. It is only when patient caretakers are empowered with this knowledge and skill-set that these patients have a fair chance at living meaningful lives.

Accurate diagnosis of DWC is the first step in treating DWC-associated hydrocephalus. Failure to do so may result in progressive hydrocephalus, cerebral cortical compression and atrophy, irreversible macrocephaly, herniation, and death. Recognition of signs and symptoms of hydrocephalus should prompt

neuroimaging to define the etiology of hydrocephalus, DWC, or otherwise. Using the criteria reported in this manuscript, we believe three entities along the DWC continuum can be accurately diagnosed using CT scanning instead of MRI. Patients can then be offered a variety of surgical treatment options with the goal of resolving hydrocephalus.

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Language of the Heart: A Student's Perspective on Congenital Heart Defects and Volunteering

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Lan Xiao Hua is a 13 year old girl from Gansu, China. She had Ventricular Septal Defect, a hole in her heart's right ventricle. Doctors said she would be able to live to approximately 20 years of age. Xiao Hua's family knew about her congenital heart disease (CHD) since she was 7 years old, but with the humble living they made coaxing wheat from the dry, parched land of Gansu Province, the necessary surgery expenses would have required 10 years' worth of their income.

Xiao Hua's condition is not untreatable. Rather, CHD requires what is considered one of the simpler cardiac surgeries. Had Xiao Hua been operated on earlier, the chances of a successful surgery would have been almost 99%. After the surgery, she would have been completely normal, with

few lingering symptoms. Instead, without the necessary surgery for CHD, Xiao Hua was easily exhausted by the simplest of everyday activities, such as walking. Her lips were tinted blue whenever she did any work. Even breathing was painful for her. Xiao Hua was more susceptible to transmittable diseases and caught colds frequently, a symptom of poor blood circulation. She was also shorter than other 13-year-old girls because her faulty heart contributed to stunted growth and development. Xiao Hua desperately needed the heart surgery, but she didn't have the resources needed to obtain treatment.

Lan Xiao Hua's plight is not unusual. Congenital heart defects (CHD) are a leading cause of child mortality both worldwide and in the U.S. According to the American