Normal Pressure Hydrocephalus

Author: Ashley Fink
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Normal pressure hydrocephalus (NPH) is a condition that is often hard to discern and differentiate from similar pathologies. This paper will explore the etiology of this pathology, its clinical manifestations and symptoms, and how NPH is treated. With knowledge of its symptoms, early detection of this pathology will allow for early intervention to greatly improve the prognosis of affected individuals.

NPH was discovered as the first treatable form of dementia in the 1960s by Hakim and his colleagues: Adams, Fisher, Ojemann, and Sweet. This condition occurs when excess cerebrospinal fluid accumulates in the ventricles, which results in ventriculomegaly, or dilation of the ventricles, to compensate for the increased pressure (see figs. 1, 2, & 3). By expanding, this brings the pressure back down within the ventricles around the normal level of 15 mmHg, hence the use of the terminology ‘normal pressure’ hydrocephalus (Kiefer and Unterberg 16).

There are two categories of hydrocephalus: communicating and non-communicating. Communicating hydrocephalus describes a “free-flow of CSF to the venous sinuses [and] the subsequent connection to the spinal cord” with “delayed or reduced absorption at the venous sinus level” (Jayamohan 101). In other words, there is no obstruction to CSF flow but there is something causing retardation of reabsorption. In non-communicating or obstructive hydrocephalus, the accumulation of CSF is the result of a blockage, as implied by the name. NPH falls into the former category, as a communicating hydrocephalus, along with secondary forms of hydrocephalus caused by trauma to the brain, head injury, subarachnoid hemorrhage, and meningitis (Billek-Sawhney and Jackson 2012). Normal pressure hydrocephalus is an idiopathic condition, meaning its cause is unknown, although it is prevalent in the elderly.

Fig. 1, 2, & 3. Hydrocephalus, Case courtesy of A. Prok Frank Galland, Radiopaedia.org, rID: 7258

Normal pressure hydrocephalus is a disease that is often misdiagnosed. The symptoms of normal pressure hydrocephalus are often hard to differentiate from other pathologies
such as Alzheimer's disease, Parkinson's disease, and various dementias that present with similar clinical manifestations. NPH can be identified when patients express the Hakim triad of "wet, wild, and wobbly" (Kiefer and Unterberg 17, Billek-Sawhney and Jackson 183). Although these three symptoms point to NPH, the expression of all three is indicative of a later state. Because early detection is essential for a better prognosis, this condition can be diagnosed with only one or two of the symptoms.

"Wet" refers to urinary incontinence. The progression of this symptom usually begins as an increased urinary frequency, followed by the sudden and urgent need to urinate, and finally the loss of control of urination or incontinence. "Wild" pertains to the characteristics of dementia. Patients or their caregivers may first notice "psychomotor slowing impaired attention and concentration, fine motor slowing and imprecision, and short-term memory impairment." As NPH progresses the symptoms exacerbate into "apathy, reduced drive, indifference, bradyphrenia [or slowed thought process], reduced speech production, and rarely, akinetic mutism" or the inability to move or talk (Kiefer and Unterberg 19). Because many forms of dementia or other degenerative neurological disorders have some of the same symptoms, it's important to run other tests to rule them out. The recommended tests are the "grooved pegboard test, Stroop test, digit span test, trail-making A/B test, and the Rey auditory-verbal learning test" (Kiefer and Unterberg 19). If a cognitive impairment is noted from these tests, NPH can be ruled out. "Wobbly" refers to a gait apraxia defined as "difficulty sequencing the required movements needed for ambulation [...] as well as uncoordinated attempts to alter course or maintain continuous movement" (Siedlecki 29). This awkward gait is known as magnetic gait or magnetic shuffle. It may begin as "dizziness, difficulty walking on a slope or stairs, and difficulty getting up from or sitting down on a chair" (Kiefer and Unterberg 18). Eventually the patient will walk as if their feet are stuck to the ground, shuffling with toes pointing outward in a wide stance. Although listed last, this is usually the first and most definitive of the triad to appear in patients.

If a patient presents with these symptoms, a CT or MRI scan of the head and other invasive diagnostic testing can be ordered and performed to make the diagnosis. In the images, "a fourth ventricle of normal size in the presence of enlarged lateral and third ventricles [...] is a finding consistent with NPH" (Kiefer and Unterberg 21). Although both modalities will show these abnormalities known to normal pressure hydrocephalus, MRI is the imaging modality of choice because its detail is ideal when imaging the central nervous system and it "can also add support for the diagnosis with CSF flow studies and MRS," which is a test that can show lactate peak that is sometimes reported with NPH (Hacking and Singh). One of these invasive diagnosis tests should also be performed: the spinal tap test, daily CSF drainage (150 to 200 ml for 2 to 7 days), or infusion tests (Kiefer and Unterberg 2012). With thorough testing, this increases the chances of an accurate diagnosis.

After diagnosis, NPH is usually treated in one of two ways: a ventricular shunt or lumbar tap tests. A ventricular shunt is the preferred form of treatment for NPH (see fig. 4). A plastic tube is inserted into the lateral ventricles to drain cerebrospinal fluid. This shunt empties into one of three anatomical spaces, listed in order of preference: peritoneum, pleural space, or a neck vein that drains into the atrium (Jayamohan 2010). There are different types of shunts available and they can be set and adjusted to drain CSF at different rates. Although this is the ideal form of treatment, many complications may occur. The surgery alone carries the risks of hemorrhage or stroke. Afterwards, there is a risk of infection due to a foreign body remaining inside the body. Other complications include internal bleeding along the tract of the shunt, seizures, and the most serious complication, a subdural hematoma. Patients and caregivers should be vigilant of "headaches, changes in vision, loss of coordination, fatigue or difficulty sleeping, as these may be signs of infection, bleeding or obstruction" (Siedlecki 32).

The second treatment that is common for NPH is serial lumbar punctures. While this is also part of the method for discerning if NPH is the diagnosis, it can also be used to treat patients that are not eligible for surgery. A perk of this is that it can be performed in any hospital setting, but this is not recommended for long term treatment.

Upon early detection and treatment, the prognosis for a patient diagnosed with NPH is good, with 70% to 90% of patients experiencing improvement (Kiefer and Unterberg 2012). The symptom usually relieved first is the magnetic gait, which is also the first to appear, but the patient may not return fully to his or her state prior to the condition. It should also be noted that shunting requires follow-up visits for proper care. It is recommended that postoperative visits occur two to three times in the first year because this is when complications are most likely to happen. These are physical exams, but follow up MRIs or CTs should be done in the first year and after any shunt revisions. Unless conditions worsen, no more imaging should be performed (Kiefer and Unterberg 2012).

Normal pressure hydrocephalus is a form of reversible dementia that is often misdiagnosed because of its close resemblance to other aging diseases seen in the elderly population. In this idiopathic condition, the lateral ventricles of the brain dilate to compensate for increased pressure, effectively returning to a normal pressure. Although the pressure reads as normal, the ventricular enlargement puts pressure on the brain. If a patient presents with the Hakim triad of "wet, wild, and wobbly," a diagnosis can be confirmed with a combination of CT or MRI imaging and other invasive diagnostic exams to rule out other pathologies. Shunt insertion is the ideal form of treatment to drain excess cerebrospinal fluid. With proper maintenance, including follow up imaging and physical examinations, a patient's quality of life has the potential to be greatly improved with the return of psychomotor function.
Works Cited


"Future Sound": Analysis of Digital Tools from the Vocaloid and Hatsune Miku Fan Communities

Author: Taylor Wiley
Subject: Social Informatics/Anthropology

Abstract

The singing synthesis software, Vocaloid, along with one of its most popular voice libraries, Hatsune Miku, has gained an Internet fan base thanks to an infrastructure of already-existing websites and the concept of "remix culture," which permits working off of others' ideas and assets to make new creations. In this paper, I examine some of the critical aspects and Ideas of the Vocaloid fandom through key readings and articles. These readings reveal that the popularity of Vocaloid is due to both this remix culture and Hatsune Miku's depiction by her creators and fans. I also discuss some key points concerning the fan base's relationship to technology. Finally, I propose ideas on what future research in this area might choose to focus.

This paper discusses the Vocaloid software, one of its most popular voice libraries, Hatsune Miku, and the fan base that has emerged primarily on the Internet focusing on both. Vocaloid, often stylized as VOCALOID, is a singing synthesis software that was initially developed as part of Jordi Bosada's dissertation project at the Pompeu Fabra University in Spain, with funding from the company Yamaha, who also owns the rights to the program. Initially released in 2004, the software was one of the first commercially sold singing synthesizers, the development of which had previously been confined to university research. While Yamaha is responsible for the development and maintenance of the software, they do not publish the voices. Instead, Yamaha licenses development tools to third-parties, who then "create a database (called a singer library) consisting of phonemes voiced by an organic body and then release that voice with a score editing version of the software that allows the user to write music that is sung by the synthesized voice" (Bell 223). Much like other musical software such as FL Studio and GarageBand, Vocaloid's editor software uses a piano-roll interface (Figure 1).

This software ignited a large fandom focused on the creation, dissemination, and enjoyment of creative works that use the software after Crypton Future Media, a Japanese music software distributor, decided to create anime-style avatars for their voice libraries, a strategy which has since been imitated by many other companies. These avatars are essentially "bodies" for the synthesized vocalists. Additionally, Crypton markets their libraries as "character vocals," a tactic that came about due to the fact that professional