normal pressure hydrocephalus:
Hydrocephalus is a group of neurological disorders in which excess cerebrospinal fluid (CSF) builds up in the ventricles of the brain [1]. The ventricles enlarge and compress the brain tissue, primarily in the periventricular regions, inducing significant neurological deficits. The compressed neurons can be functionally and/or structurally compromised. Gradual ventricular dilatation is initially tolerated as the local shape of the cerebral ventricles changes from a narrow shape to a more rounded shape. At some point, the surface area begins to enlarge and the local tissue stretching becomes problematic. This is associated with reductions in cerebral blood flow, emergence of oedema and the appearance of neuronal dysfunction. In case of prolonged brain compression, impairments become permanent due to destruction of axons, neuronal cell loss and tissue atrophy [2].

Normal Pressure Hydrocephalus (NPH) is a subtype of hydrocephalus that usually occurs in elderly adults [3, 4]. NPH is characterized by a slow dilatation of the ventricles but intracranial pressure remains normal or just slightly elevated with a clinical triad of gait disturbance, dementia and incontinence [4, 5]. To this day, it remains a mystery to scientists how the ventricles can dilate when CSF pressure appears to be normal, and there is no significant pressure difference between the ventricles and the outer edge of the brain. The most widely accepted hypothesis is that transient intracranial pressure peaks lead to chronic mechanical stress on ventricular walls, finally resulting in ventricular dilatation and brain compression [6]. The fluid-like viscoelastic nature of brain tissue facilitates this expansion over time. The prevalence of NPH remains uncertain, with reported rates varying from 1.3 per million to 4 per 1000, depending on diagnostic criteria for NPH and populations sampled. Recent estimates quote an annual incidence of 1.8 per 100,000 [7].

Significance of the problem – Difficulties in diagnosing the cause of dilated ventricles:
Patients with dilated ventricles are not all suffering from hydrocephalus. Other diseases such as Alzheimer’s and Parkinson’s diseases can also induce a dilatation of the ventricles and create similar symptoms (gait difficulties, dementia and incontinence). Here, the mechanism of ventricular dilatation is associated with reduced brain volume due to tissue atrophy, allowing the ventricles to dilate and fill with CSF. The similarities in the symptoms and the abnormal size of the ventricles between NPH and other disorders makes NPH patients difficult to identify, leading to misdiagnosis. This is a real challenge for the medical community[8]. It is extremely difficult to differentiate them on the basis of the neurological deficits and measuring ventricular size using conventional medical imaging techniques [8, 9]. The distinction is clinically important because NPH is one of the few treatable causes of dementia [10, 11]. The treatment of NPH consists of surgically diverting CSF from the ventricles into the subarachnoid space (third ventriculostomy) or from the cranium to the abdominal cavity via a shunt, allowing the ventricles and brain to return to normal size. Draining CSF from the dilated ventricles usually results in clinical improvements. After treatment, clinical improvements have been reported in 30-96% of patients [12]. It was found that 93% of patients have gait improvements and more than 50% have significant cognitive
improvements [13-16]. The prognosis for hydrocephalus patients depends on the cause, extent of symptoms, and the timeliness of diagnosis and treatment. In general, the earlier hydrocephalus is diagnosed, the better the chance for successful treatment [3, 17, 18]. Treatment, by removing excess CSF at the early stage of hydrocephalus development, can restore neural function. Whereas the longer hydrocephalus goes untreated, more of the neurons die under the effect of the brain compression and are unable to be rescued by treatment.

It is estimated that between 1.6 and 5.4% of patients with dementia actually have NPH [19, 20]. These people could have been treated and their symptoms improved if the NPH was correctly diagnosed. Additionally, conventional hydrocephalus treatments have no benefit in the non-hydrocephalus patients. If non-hydrocephalic patients are misdiagnosed as hydrocephalic patients, treatment have been shown not only to be ineffective, but are associated with significant morbidity[21]. Since the overall prevalence of dementia rises progressively as the population ages, even if NPH is responsible for only a small proportion of senile dementia, successful treatment could help large numbers of patients. The challenge in the elderly population is sorting through changes that result from normal aging and those comorbid illnesses commonly encountered when making this determination [22]. Improving the diagnostic criteria to differentiate NPH patients from other diseases with ventricular dilatation has the potential to reduce medical costs by USD180 million/annum in United States alone[23].

The current contribution of medical imaging:
Computed tomography (CT) and magnetic resonance imaging (MRI) depict ventricle size accurately. However, currently, findings on brain images are not sufficient on their own to establish a diagnosis because they provide minimal, if any, direct evidence of brain damage despite marked deficits in motor skills and cognitive functioning. Other methods including single-photon emission CT, positron emission tomography, nuclear cisternography and CSF flow velocity were trialed in NPH diagnosis, but have unclear diagnostic value. At present, these examinations are not part of the routine work-up of patients with suspected NPH [10, 24, 25].

References:


