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Could hydrocephalus shunts have a role in the treatment of lymphoedema?

Bell J & Piller N

ABSTRACT

Lymphoedema is a consequence of impaired lymphatic drainage. Available treatment options vary in efficacy and impact on the individual. Whilst all are useful in reducing the extent and impact of lymphoedema, there are confounding factors such as patient compliance, financial and physical costs and unpredictably variable outcomes. There seems to be no single treatment that is affordable, effective and sustainable for patients with lymphoedema.

This review introduces the novel idea of a modified hydrocephalus shunt as a surgical alternative to treat (and perhaps prevent) lymphoedema.

Hydrocephalus shunts allow cerebrospinal fluid (CSF) to circumvent an obstruction during periods of impaired absorption, removing the build-up of fluid that causes hydrocephalus, working on a low pressure system.

Lymphatic pumping pressures in failing arm lymphatics have been recorded as approximately 25 mmHg¹. In early stage leg lymphoedema, pressures have been recorded around 70mmHg (diastolic) and 100mmHg (systolic), whilst late stage lymphoedema has been recorded as 20mmHg, though often these lymphatics are very difficult to cannulate.

As hydrocephalus shunts work on pressures as low as 15-25mmHg at flow rates as low as 5ml/hr it is plausible that they could be used to facilitate normal flow, but

importantly prevent retrograde flow of lymph in failing or failed lymphatics, thereby reducing lymphoedema.

Keywords: *Lymphoedema, hydrocephalus, shunt, intralymphatic pressure.*

INTRODUCTION

The lymphatic system is a low pressure vascular network that has a major role in the maintenance of fluid homeostasis and immune system regulation and defence in the human body² and is often at risk of damage through illness, surgery, radiotherapy, infection and day-to-day life.

There is a net, low-level filtration of plasma from both the arterial and venous ends of capillary beds, resulting in accumulation of plasma in the interstitium³. This ultra-filtrate comprises fluid, proteins and small molecules which are able to filter out of the capillaries and into the interstitium⁴ and this interstitial fluid is then taken up into the lymphatic system by the terminal lymphatic capillaries⁵.

Lymphoedema is a process of either congenital (primary lymphoedema) or acquired (secondary lymphoedema) abnormalities of the lymph vessels that result in impaired lymphatic drainage and fluid accumulation in the tissues^{6,7}. Damage to lymphatic vessels can result in degenerative changes such as hyperplasia of endothelial cells, thickening of sub-endothelial collagen fibres, decreased number of collagen fibres and eventual total occlusion of the lymphatic lumen⁸. These changes result in an initial deceleration of flow, which causes overloading of the lymphatics and increased intra-lymphatic pressure with subsequent dilation and incompetency of unidirectional valves and retrograde lymph flow. Eventually, due to lack of contractility of lymphatics there are no effective pressures produced by the lymphatics sufficient to propel lymph and as the outflow for lymph is blocked, muscle contractions are unable to empty excess lymph⁸. This reduction in lymph flow often predisposes patients to poor wound healing, causing further complications.

In developed countries, the greatest cause of lymphoedema is obstructive lymphoedema caused mainly from the treatment of cancer; such as those patients who have received mastectomies and lumpectomies⁶. Outside of developed nations the most common cause of lymphoedema is lymphatic filariasis, which affects approximately 120 million

Jemima Bell*

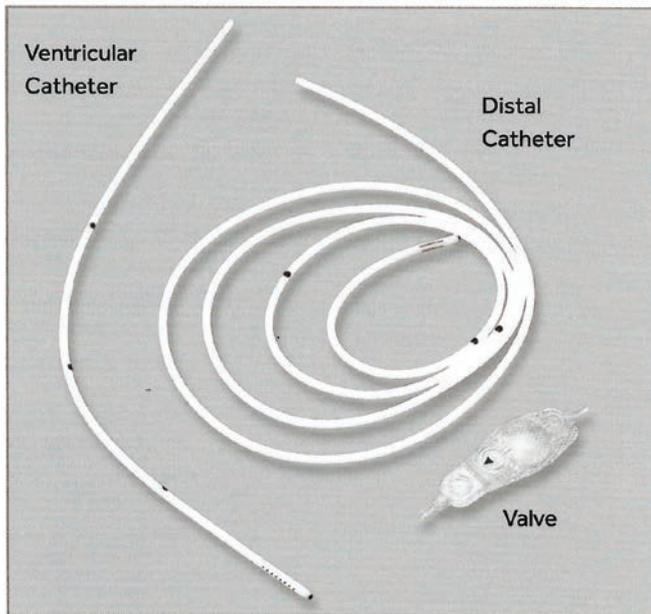
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Figure 1: Main components of a hydrocephalus shunt¹⁴



people worldwide^{6,9}. There are functional, psychosocial and emotional aspects of lymphoedema which impact greatly on the quality of life of sufferers, some of which are associated with the treatment regimens prescribed¹⁰.

CURRENT TREATMENT OPTIONS

As with most interventions, a multidisciplinary approach to the treatment and management of lymphoedema is most effective, currently referred to as complete decongestive physiotherapy or treatment^{6,11}. For lymphoedema, current research dictates that an approach that involves exercise and movement, swelling reduction and maintenance, skin care, risk avoidance and management of pain and psychosocial issues offers the best outcomes for lymphoedema sufferers⁶. There is currently an array of non-surgical and surgical treatment options available that offer varying degrees of relief for patients.

Physical therapy combined with compression is considered the standard treatment aiming to reduce fluid outflow into the interstitium by reducing venous pressure and flow¹¹. This has been shown to reduce the size of the oedema by 40–60%, with compression garments achieving a size reduction of 46% individually⁶. However, this treatment is merely a maintenance treatment, not a cure, and one which relies heavily on long-term patient participation and compliance⁶.

Manual lymphatic drainage is a form of massage developed with the aim of directing fluid towards functioning lymphatics and thereby bypassing damaged or obstructed lymphatics in order to drain an area of congestion⁶. Extensive studies have demonstrated that it is not beneficial as a stand-alone treatment, and that if done inappropriately (that is, too forcefully) can actually damage lymphatic vessels and

increase capillary filtration, which will only worsen oedema. As such, it is a treatment that should only be performed by trained operators, be that therapists, caregivers or the patients themselves^{6,11}.

Pneumomassage, or intermittent pneumatic compression (IPC) involves compression supplied by an external pump and compression stockings to reduce oedema⁶. This form of intervention is limited by patient compliance, risk of lymph obstruction, and lack of suitability for those with renal or congestive heart failure, and active metastatic disease. There is also a risk of damage to more superficial structures if the pressures in the pump are set too high⁶.

Liposuction in the treatment of lymphoedema aims to remove fat in the affected limb in order to reduce the appearance and symptoms of lymphoedema⁶. It is indicated when other, non-surgical treatment options have been exhausted and there is significant discomfort in the affected limb due to weight¹². It is, however, not a treatment option for patients with pitting oedema and requires high levels of patient compliance post-surgery with the use of compression garments and wound care⁶.

Anastomoses can either be lymphatic to lymphatic (lymphatic-lymphatic bypass), lymphatic to vein (lymphaticovenous bypass) or lymph node to veins and arteries (microvascular lymph node transfer)⁶. These interventions aim to bypass the obstructed lymphatic. Whilst these surgical interventions have been shown to reduce lymph clearance time, there are potential complications, such as the development of lymphoedema at the site of lymphatic harvest and large surgical scars, combined with the fact that there is limited standardised data on the efficacy of such interventions⁶.

HYDROCEPHALUS SHUNTS

Hydrocephalus shunts traditionally allow cerebrospinal fluid (CSF) to circumvent an obstruction in its normal pathway during periods of impaired absorption, removing the build-up of CSF that causes hydrocephalus. These shunts usually comprise three components: an inflow catheter which drains CSF directly from the ventricles or subarachnoid space; a valve which regulates flow and pressure within the catheter; and an outflow catheter, which connects either to the peritoneal cavity, heart or other drainage site and allows CSF to flow out into this space (Figure 1)¹³.

In order to assess whether the use of hydrocephalus shunts would be appropriate in the treatment of lymphoedema, two brands of hydrocephalus shunts commonly used in Australia were considered: Medtronic[®] and Codman[®]. Both companies boast an array of shunt systems with catheters designed for various specifications and valves that may operate under single or adjustable pressures^{15–17}. Comparing the pressure-flow rate settings of the Codman CERTAS[®] Plus Programmable valve and the Medtronic[™] Strata[™] adjustable valves it can be seen that both operate similarly under very low pressures and flow rates (Figure 2)^{15,17}.

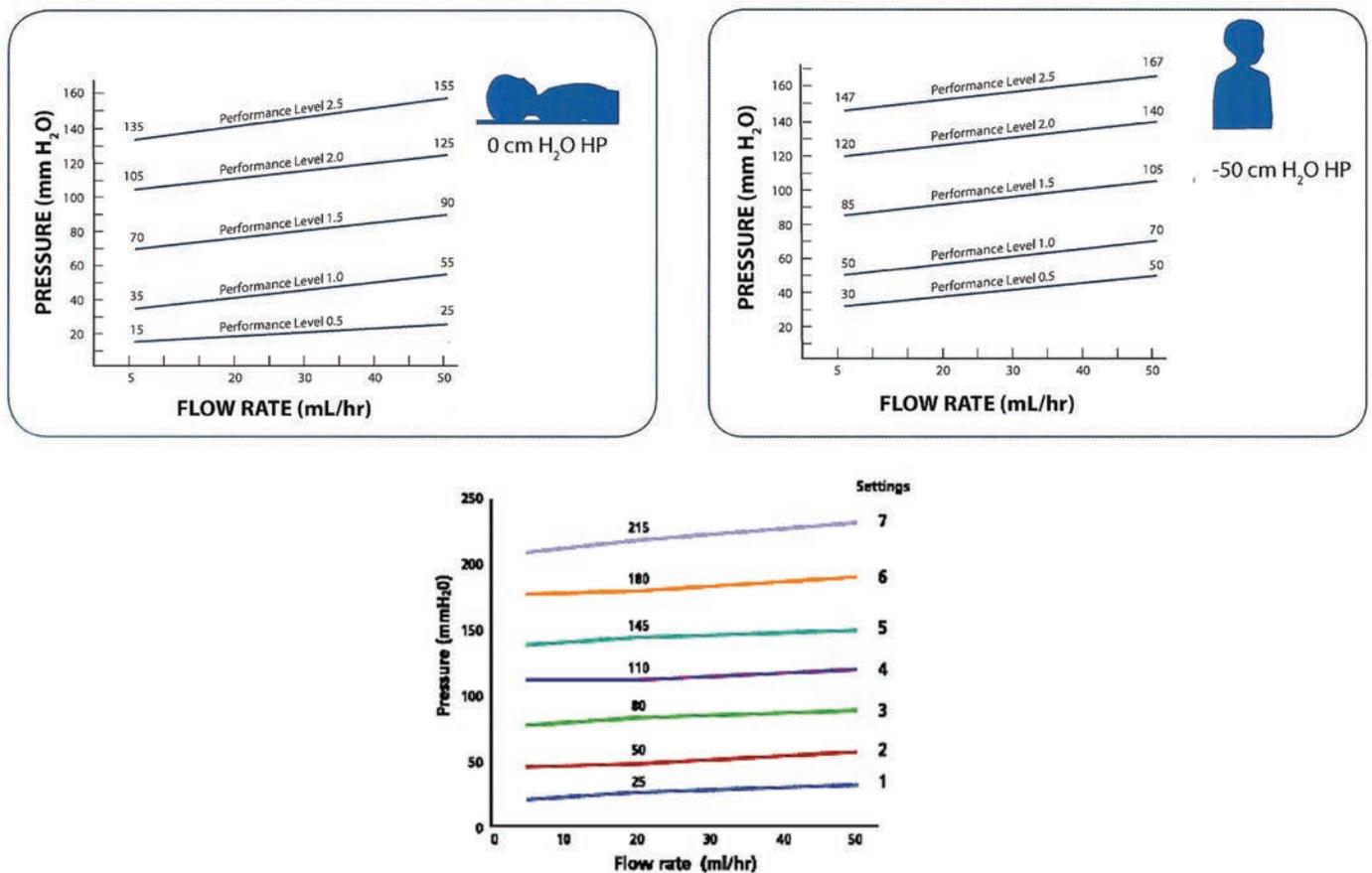


Figure 2: Top are the specifications for the Medtronic™ Strata™ adjustable valves and below are the settings for the Codman® CERTAS® Plus Programmable valve^{15,17}.

As demonstrated by Figure 2, these adjustable valves operate under pressures as low as 15 mmHg and flow rates as low as 5 mL/min^{15,17}.

HYDROCEPHALUS SHUNTS AND INTRALYMPHATIC PRESSURES

There is limited data available with regard to the pressures acting within human lymphatics; however, studies by Modi *et al.* and Olszewski and Engeset have shed some light on lymphatic flow rates and pressures under various circumstances. In this review, end lymphatic pressures are being focussed on, as this data will be most relevant to the potential use of a hydrocephalus shunt as treatment.

It is important to note that in healthy individuals, the rhythmic contractions of the lymphatics are the main factor in the generation of pressures sufficient to produce lymph flow, and therefore contractions of surrounding musculature, and external massage and compression of the lymphatics of healthy individuals do not greatly increase flow as the lymphatics are usually empty between pulsations^{8,18,19} and the pulsations alone are sufficient to propel lymph. Whilst rhythmic contractions of surrounding leg muscles do not increase the mean pressure of normal lymphatics

between pulsations, it does create a slight increase in pulse pressure of 0.5–3 mmHg superimposed on the intrinsic pressures produced in both horizontal and upright healthy patients^{8,20}. These contractions also increase the frequency of pulsations^{8,18,20}.

In individuals with obstructive lymphoedema, spontaneous contractions are irregular and ineffective, whilst muscle contractions, external compression of lymphatics through the use of elastic stockings and massage can produce lymph flow¹⁹.

Several studies have demonstrated that a systolic pumping pressure of 5–25 mmHg in normal leg lymphatics is required for lymphatic pulsations to begin to occur^{8,18,20}, with most healthy lymphatics reaching systolic pumping pressures of 29–37 mmHg, sufficient to propel lymph, with a tendency towards higher pressures in larger vessels^{8,20}. Diastolic pressures were usually around 0 mmHg between pulsations, during which time there is no flow of lymph^{8,18,20}. In the upright position, normal leg systolic pressures were approximately 44.7 mmHg^{8,20}; however, there have been other studies that have recorded no significant difference between supine and standing healthy leg pressures²⁰.