CEREBROSPINAL FLUID LEAKS IN RELATION TO EAR, NOSE & THROAT

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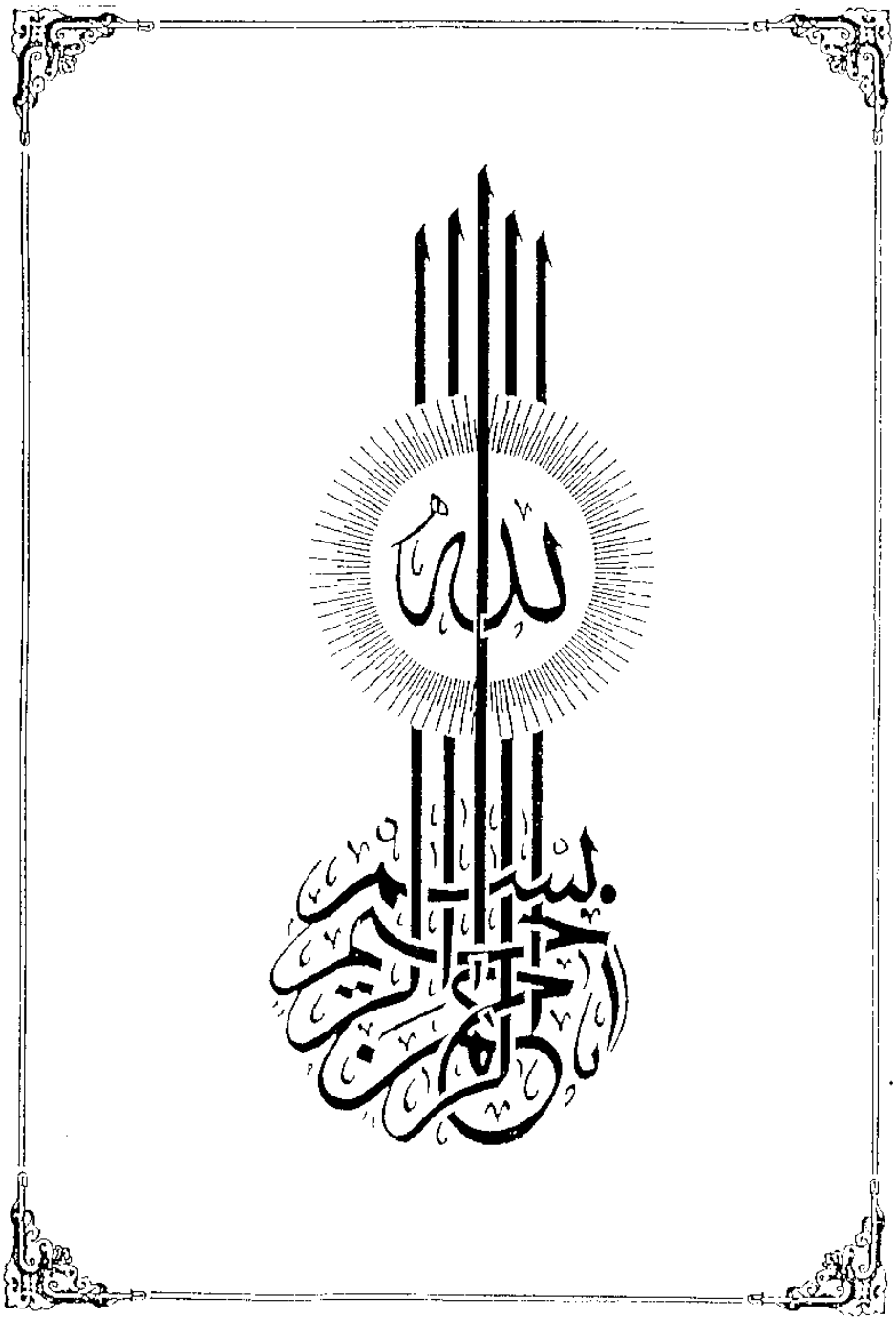

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INTRODUCTION

Cerebrospinal fluid leakage is one of the problems facing neurosurgeons and otolaryngologists, due to its occasional difficulty in diagnosis, localization and treatment, as well as due to the catastrophic significance of ensuing infection *(Pomeranz et al., 1991)*.

Leakage of CSF occurs when the arachnoid, dura, bone and epithelium are violated resulting in extracranial flow of CSF. This occurs due to either congenital malformations, or, more commonly, as acquired forms of CSF fistulae accounting for 90% of cases, and occurring secondary to surgery, infections, tumours, or, most commonly, accidental trauma *(Brodsky, 1984; Applebaum and Chow, 1993)*.

The CSF leak may present in various forms: rhinorrhea, otorrhea, otorhinorrhea, serous otitis media *(Seligman and Lusk, 1988)*, operative wound leak, CSF leak from a compound fracture of the cranial vault, or in the form of recurring meningitis *(Raaf, 1967; Myers and Sataloff, 1984)*. In most reported series of patients, **CSF rhinorrhea** is the more common presentation followed by CSF otorrhea *(Applebaum and Chow, 1993)*.

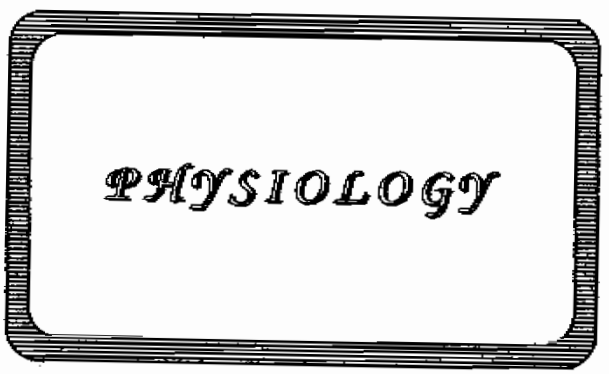
Physicians may overlook the leakage in a trauma patient with other serious injuries, or confuse the clear fluid discharge with other pathologic conditions, thus subjecting the patient to the high risk of severe recurring infectious meningitis that occurs with CSF fistulae *(Mamo et al., 1982; Applebaum and Chow, 1993)*.

Meningitis occurs in approximately **24%** of patients presenting with CSF leaks *(Brodsky, 1984)*; and the most commonly isolated organism in such cases is

Pneumococcus, followed by **Hemophilus influenzae** *(Papay et al., 1989)*.

Meningitis occurring with CSF leaks has an appreciable morbidity and mortality *(Calcaterra and Rand, 1973)*. **20%** of children having a fistula between the middle ear and subarachnoid space will have partial or complete sensorineural hearing loss after each attack of meningitis, and this incidence increases with recurrent meningitis *(Kaseff et al., 1980)*. The mortality rate in patients suffering from meningitis with CSF leaks is as high as **20%** in the antibiotic era *(Fishman, 1980)*, and increases to **30-50%** in cases of recurrent meningitis caused by a persistent CSF fistula (a fistula that persists for more than two weeks) *(Chandler, 1983; Loew et al.,1984; Hubbard et al., 1985)*.

The aim of this paper is to review in a detailed manner the documented literature concerning the aetiology, diagnosis and management of CSF leaks in relation to ear, nose and throat including the most recent and up-to-date references available.


PHYSIOLOGY

PHYSIOLOGICAL CONSIDERATIONS

CSF CIRCULATION:

Spinal fluid is produced by the **choroid plexus**, which is a cauliflower growth of blood vessels covered by a thin coat of epithelial cells and projects into the lateral, third and fourth ventricles; CSF continually exudes from the surface of the choroid plexus into the ventricles *(Guyton, 1977)*. This occurs in a manner much like that of urine production by the glomerulus *(Myers and Sataloff, 1984)*.

However, *Ganong (1991)* estimates that the choroid plexuses account for **50%** to **70%** of the CSF produced, and the remaining is being formed around the cerebral vessels and along the ventricular walls.

CSF passes from the lateral ventricles through the foramen of **Munro** into the third ventricle, through the **aqueduct of Sylvius** into the fourth ventricle, from which CSF reaches the subarachnoid space via two foraminae adjacent to the cerebellum - the foramen of **Magendie** medially and the foramen of **Luschka** laterally. It circulates down around the spinal cord and up around the basal cisterns and over the surface of the brain to be absorbed into the blood of the dural venous sinuses through the arachnoid granulations *(Myers and Sataloff, 1984)*.

These granulations are arachnoid trabeculae that protrude from the subarachnoid space through the venous walls, resulting in extremely permeable areas that allow relatively free flow of CSF into the blood. The reabsorption of CSF occurs by a process of **pinocytosis** involving active encapsulation of the fluid into intracellular packets, transport across the cell itself, and discharge of the packet into the superior sagittal sinus. The process is quite sensitive and there is a fairly narrow margin for accommodating additional CSF. Additional spinal fluid is secreted by the brain itself, and this may account for as much as one third of CSF volume, depending on the circumstances *(Myers and Sataloff, 1984)*.

CSF PRESSURE:

In adults, the pressure in the lumbar subarachnoid space with the patient lying on one side is normally **50-150 mm of CSF** *(Macleod et al., 1988). Ganong (1991)* estimates this pressure to be **70-180 mm of CSF**, averaging **112 mm CSF**. In infants, normal CSF pressure is about **40 mm CSF** *(Beckhardt et al., 1991).*

The head position, as well as the normal arterial and respiratory pulse waves affect the intracranial CSF pressure *(Calcaterra, 1980)*, and it is subject to recurring fluctuations averaging **80 mm CSF** every few seconds *(O'Connell, 1964).*

RELATION BETWEEN CSF PRODUCTION AND VOLUME:

The amount of CSF produced daily from the choroid and the brain is approximately **500 ml** *(Pearson, 1991).* The rate of production is about **0.3** c.c. per minute *(Myers and Sataloff, 1984).* However, the volume of CSF in a normal adult is between **90 and 150 ml**. Therefore, one could leak more than **300 ml** of CSF each day and still maintain an adequate volume for normal functions *(Pearson, 1991).*

FACTORS AFFECTING CSF PRODUCTION AND ABSORPTION:

Medications such as Acetazolamide **(Diamox)**, Frusemide **(Lasix)** and **Digitalis** can substantially reduce the CSF production rate *(Myers and Sataloff, 1984)*; while increased production of CSF may result from overdevelopment of the choroid plexus in the infant *(Guyton, 1977).*

However, the rate of CSF production is fairly independent of intraventricular pressure, blood pressure and other physiological factors, unlike the **absorption** of CSF, which takes place largely by bulk flow, and is proportionate to the intracranial CSF pressure. Below a pressure of approximately **68 mm CSF**, absorption stops to maintain adequate CSF volume for normal functions. At **112 mm CSF**, absorption rate equals the production rate. Above a pressure of 112 mm

CSF, the absorption rate exceeds the production rate, and gradually increases with increase in the CSF pressure *(Ganong, 1991)*.

Large amounts of fluid accumulate when the reabsorptive capacity of the arachnoid villi is decreased **(external, communicating hydrocephalus)**, as in case of *thrombosis of the sagittal sinus*, or due to interference with pinocytosis by *hemoglobin breakdown by-products*. Fluid may also accumulate in the ventricles proximal to a block **(internal, noncommunicating hydrocephalus)**; the block may either be in the foraminae of Luschka and Magendie (as by *inflammatory exudate*), or in the ventricular system (as in case of *intraventricular tumour* or from *extrinsic compression* by a space-occupying lesion or oedema) *(Myers and Sataloff, 1984; Macleod et al., 1988; Ganong, 1991)*.

CHEMICAL COMPOSITION OF CSF:

The formation of CSF depends on filtration and diffusion from the blood, along with facilitated diffusion and active transport mainly across the choroid plexus. The composition is essentially the same as that of the brain extracellular fluid, and there appears to be free communication between the latter, the ventricles and the subarachnoid space. The blood-brain barrier governs the exchange of substances across capillary walls between the plasma in the cerebral blood vessels and choroid plexus on one hand, and the extracellular fluid and CSF on the other hand. Different substances have different rates of penetration into the brain and CSF. This accounts for the difference in composition of plasma and CSF ***(Table 1)***. Drugs given in managing diseases of the nervous system should be able to cross to the CSF and brain. Among antibiotics, **sulfadiazine** and **erythromycin** readily cross the blood-brain barrier unlike penicillin and chlortetracycline *(Ganong, 1991)*.

The blood-brain barrier breaks down in areas of the brain that are **irradiated**, **infected** or the site of **tumour**. Substances like radioactive iodine-labeled albumin penetrate normal brain tissue very slowly, but they enter rapidly into areas with

broken-down blood brain-barrier as tumours, thus helping in their localization. Sudden marked increases in blood pressure or intravenous injection of hypertonic fluids can lead to temporary disruption of the barrier *(Ganong, 1991)*.

FUNCTION OF CSF:

This is a matter of conjencture; it probably acts as a **mechanical cushion** for the brain. The brain is supported within the arachnoid by the blood vessels, nerve roots and multiple fine fibrous arachnoid trabeculae running in the subarachnoid space. The arachnoid is held to the dura by the surface tension of the thin layer of fluid between the two membranes - there is normally no subdural space. The dura is attached firmly to bone. In case of a blow to the head, the arachnoid slides on the dura and the entire brain moves simultaneously, because the brain and CSF have approximately the same specific gravity, so that the brain simply floats in the fluid, thus decreasing its weight from **1400 grams** in air to **50 grams** in CSF. The ability of the brain to float in the CSF permits its relatively weak attachments to suspend it effectively *(Guyton, 1977; Ganong, 1991)*.

Furthermore, the CSF seems to act as a **metabolic sink**. There are no lymphatics in the brain and metabolites diffuse into CSF, ultimately ending in the blood stream *(Pearson, 1991)*.

The pain and severe headache by spinal fluid deficiency during, for example, pneumoencephalography, illustrate the importance of CSF in preventing the brain from hanging on the vessels and nerve roots, which would lead to traction on them and stimulation of pain fibres *(Ganong, 1991)*.

CSF-PERILYMPH RELATIONSHIPS:

Communication of CSF and perilymph in humans has been proven by many investigators *(Arnvig, 1951; Igarashi and Schuknecht, 1962; Rice and Waggoner, 1966)*. The most crucial CSF-perilymph relationships function either through the

internal auditory canal neural foramina, or through the **cochlear aqueduct** *(Goodhill, 1981)*. Variations in the patency of both these connections between the CSF and perilymph can be significant *(Tonkin and Fagan, 1975)*, ***(Fig. 1)***.

The cochlear aqueduct (CA) becomes longer and relatively narrower with age; it is believed to be functionally patent in most adult persons, and fluids intermingle through it by diffusion. However, in most individuals, the CA can neither transmit sudden large pressure changes to the inner ear, nor allow free flow of CSF in any significant quantity; this is because the CA is narrow and contains a fibrous tissue meshwork occupying and baffling its lumen *(Palva and Dammert, 1969; Jackler, 1993)*.

An abnormally patent CSF-perilymphatic communication can lead to a CSF leak into the middle ear during surgery on the stapedial footplate (where a **gusher** occurs), or can result in a **non-traumatic, spontaneous CSF leak** into the middle ear *(Chandler, 1983; Ohlms et al., 1990)*. Such details will be discussed later with "CSF Otorrhea and Otorhinorrhea". The exact location of an abnormal anatomic CSF-perilymphatics communication is controversial *(Holden and Schuknecht, 1968)*. However, there are two commonly acknowledged connections:

A. The first is the presence of a **widened, patent cochlear aqueduct**, in which there is less than normal or even absent fibrous tissue; this allows CSF to pass directly into the scala tympani, through the helicotrema and scala vestibuli to the vestibule *(Freeland, 1973; Schuknecht, 1974; Jackler, 1993)*. There are reports of human temporal bones with abnormally patent cochlear aqueduct *(Schuknecht and Seifi, 1963)*.

B. The second possible route is an abnormal communication between the fundus of the **internal auditory canal** and the inner ear vestibule *(Skolnik and Ferrer, 1959; Glasscock, 1973; Clark et al., 1978)*. Embryologically, the internal auditory