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Living with Superior Semicircular Canal Dehiscence Syndrome

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I have bilateral Superior Semicircular Canal Dehiscence Syndrome (SCDS). What on earth is that, you may well be asking, as so few people have heard of it? It is commonly referred to as a balance disorder, but for the purpose of this post, it is essential for me to emphasise that there are people who have SCDS who have hearing loss only, and no vestibular symptoms.

To summarise, with SCDS the patient can:

1. be completely asymptomatic
2. have vestibular symptoms only
3. have hearing loss only
4. have autophony or tinnitus only
5. have any combination of 2. to 4.

I fell into the last category, but the important point to take on board is that the presentation is very varied from one person to the next.
The syndrome was discovered in 1995, by Dr Lloyd Minor and his team at the John Hopkins University School of Medicine in Baltimore, and for years it was thought to be a very rare condition. However, latest estimates in 2016 demonstrate a significant increase to show it may not be so rare after all.

The developmental anomaly occurs during the first three years of life and is thought to result from a lack of bone growth in the uppermost balance canal of the inner ear, which can cause a tiny hole to develop. In speaking to the ear and balance specialists in America that have most experience with SCDS patients, consensus suggests approximately 50% of cases as bilateral, which includes my own - in other words, on both sides of the head. Very few children exhibit symptoms, even though the rate of finding SCDS on a CT scan is the same for adults and children. Dr Gerard Gianoli at the Ear and Balance Institute in Covington, Louisiana became my surgeon after an unsuccessful operation in England. He is one of the world’s leading specialists for the condition. He suspects that all SCDS people are asymptomatic until a second event, such as a pressure altering occurrence, or trauma - this could be direct such as a blow to the head, or barotrauma as happened to me on a flight.
‘The older we get,’ says Dr Gianoli, ‘the more lax the round and oval windows inside the ear become from the repeated pressure strains. I suspect the windows get stretched to a critical point where there is increased compliance in the inner ear, and then the entire ear “jiggles” with pressure. Not everyone recognises the second event. It could just be the straw that breaks the camel’s back –

something that seems innocuous but creates the last bit of pressure to cause the breakdown.’

The problem event for me was a sharp change of pressure during the descent at the end of a flight. At the time it happened, I had a brutally sharp pain in my left ear. Going upwards in a plane is almost never a major problem for the ears as the flow of air, while the pressure in the middle ear is equalised, is from the middle ear out through the Eustachian tube – a little like a balloon deflating. Coming down or landing is harder, as the deflated balloon has to re-inflate. If the pressure change is rapid, that task of re-inflating becomes even harder.

Over a period of months after the flight more and more symptoms crept in, and they were subtly changing or worsening during that time. The headaches were becoming severe, the nausea persistent, my vision was often blurred, and it was extremely uncomfortable to raise my voice, sing or even eat crunchy foods. All the sounds reverberated inside my head. Autophony is literally defined as hearing self. More broadly interpreted, it refers not just to hearing your own voice loudly but also other internal noises, such as heartbeat and joint movements.

Furthermore, I had great trouble hearing people in noisy surroundings, and yet I was also hypersensitive to certain sounds such as a banging door, my dog barking or a spoon clattering onto the floor. I became increasingly aware of creaking noises from my neck and the clicking sounds of my eyes, and as time progressed I also had quite severe tinnitus – terrible whooshing and thumping noises in my ear. In my ignorance, I had associated the word tinnitus with ringing in the ears. But of course tinnitus takes many forms and the sound of my heartbeat pounding was causing the thumpings, and a change in blood flow near the ear, the swooshings. This is called pulsatile tinnitus.

The vertigo eventually became debilitating as I would have frequent freeze-on-the-spot attacks in certain types of environment, and it was becoming dangerous to drive. I was no longer just unsteady, everything was starting to spin. Not surprisingly, all these symptoms had a knock-on effect on my brain. My memory and concentration were noticeably affected. I had no idea at that point that my years and years of imbalance and associated anxiety were directly linked to the new symptoms that developed after the flight. All I knew was that something was making my life unbearable, and as I had a severely disabled child to care for as well, I no longer felt able to cope.

There is a general lack of awareness about SCDS among doctors. Many of those who have heard of it tend to think that they are unlikely to ever encounter a patient themselves because it’s rare. But they are failing to take into account the fact that, since its discovery just over twenty years ago, the data has been slowly accumulating and it’s not nearly as uncommon as first thought. Cases are almost certainly being regularly overlooked or misdiagnosed which is extremely concerning. It took me nearly four years to get my diagnosis and they were utterly miserable years. I only got to the bottom of my symptoms by constantly researching, and writing to different kinds of experts around the world for their advice about what might be wrong with me. Had I not done so, I simply never would have been diagnosed.

Even when I was eventually given a diagnosis, it proved to be an incomplete one. It later turned out that I had bilateral SCDS, not unilateral, and more was wrong with my left ear. Subjects with SCDS are more susceptible to a perilymph fistula (PLF). In between the air-filled middle ear and the fluid-filled space of the inner ear, very small, thin membranes act as little dividers – they are called the oval and round windows. A tear in one, or both, is called a PLF and the invisible damage also has very debilitating symptoms.

The physicians I encountered in consultations were at times brusque, and several clearly didn’t believe there was anything wrong enough to pursue. In their eyes I seemed to be a relatively fit and high-functioning person, whereas the way I saw it, my life was slowly disintegrating. A hidden condition such as SCDS is very isolating indeed. I began to question my mental state as I was being made to feel that my symptoms couldn’t be as bad as I was making out, because no-one could find anything very wrong with me. Some high tone, sensorineural hearing loss had been found in my right ear but none of the ENT specialists could account for why it had occurred. My American surgeon later confirmed, was almost certainly a result of the hole on that side of my head, and the hearing could possibly worsen if the hole wasn’t surgically repaired.
My whole experience with SCDS and related symptoms has now been published in a memoir entitled A Hole in My Life. Within the narrative I have woven a considerable amount of factual information, and it has all been authenticated by two ear and balance specialists in Louisiana.

SCDS is a condition that every audiologist needs to be aware of. There are clues that an audiologist can watch out for in an audiogram and they might be the first signs pointing towards a diagnosis. Everyone needs to bear in mind that the symptoms of SCDS can sometimes be subtle and therefore easily missed, and there is also overlap with other disorders such as hyperacusis, patulous Eustachian tube and PLF. It is extremely important to detect SCDS as it is potentially repairable with surgery, further hearing loss may be prevented and the person's quality of life can be dramatically improved.