Superior semicircular canal dehiscence (SSCD) has been defined as the absence of bone overlying the superior semicircular canal facing toward the dura of the middle cranial fossa. SSCD has been implicated as the cause of a variety of inner ear symptoms including Tullio's phenomenon, pressure induced vertigo, aural fullness, autophony, conductive hearing loss, and fluctuating or progressive sensorineural hearing loss. Additionally, SSCD has also been reported to be asymptomatic. In the past, many patients with SSCD had been misdiagnosed as having otosclerosis, patulous eustachian tubes, middle ear perilymphatic fistulas, or Ménière's disease. Identification of this entity requires a high degree of suspicion, appropriate findings on physical exam, lab testing, and confirmation on high-resolution CT scan. Surgical repair of the SSCD or occlusion of the superior canal has been reported with a high degree of symptom resolution. SSCD has been labelled the "great otologic mimicker" because it can simulate the symptoms of so many other ear disorders. However, the most common symptoms are vertigo/dizziness elicited by pressure altering activity, Tullio's phenomenon (sound-induced vertigo), fullness/pressure in the ear and autophony. Autophony is hearing internal noises louder than would be expected, such as hearing your eyes move/blink, heart beat or joint movements. Hearing loss and fluctuating hearing loss can occur, mimicking otosclerosis or Meniere's disease. Fullness in the ear and hearing your breathing loudly in the ear are symptoms of patulous eustachian tube, but can also be found with SSCD. Finally, most patients with the anatomic defect of superior semicircular canal dehiscence have no symptoms at all for some time prior to developing symptoms. SSCD can affect all age groups. There are no studies looking at the demographics of SSCD, however, the vast majority of patients diagnosed with SSCD are adults. Although the incidence of finding SSCD on a scan is the same for adults and children, the incidence of symptomatic SSCD in children is much less. Dehiscence of bone in other parts of the otic capsule can produce similar syndromes, although they are much less common. Other locations include the posterior semicircular canal, the horizontal semicircular canal and cochlea. Posterior semicircular canal dehiscence is often due to a high riding jugular bulb but can also been seen with anomalies of the vestibular aqueduct, such as enlarged vestibular aqueduct syndrome (EVAS). EVAS, by itself can also produce symptoms similar to SSCD. Horizontal semicircular canal dehiscence is most frequently the result of an eroding process such as cholesteatoma. The cochlea can have bony dehiscence where it abuts the carotid artery and at the labyrinthine segment of the facial nerve. Lastly, the bony opening where the hearing nerve enters the cochlea (at the internal auditory canal) can be abnormally wide, resulting in symptoms similar to SSCD. Because the condition of SSCD is still fairly new (identified in 1998), it could be argued that all treatments are considered experimental or investigational. However, the most frequently employed are surgical. Surgery for SSCD has included resurfacing the defect with a variety of materials, plugging the superior semicircular canal, and a combination of resurfacing and plugging. These techniques can be done through a transmastoid or a middle fossa craniotomy approach. These techniques have found great success in reducing or eliminating the vestibular symptoms of SSCD. These techniques are also effective in reducing or eliminating autophony but they have not been effective in improving hearing. A lesser approach that seems to give similar benefits is reinforcement of the middle ear windows. This surgery has the advantage of being a more minimally invasive procedure but the success rate seems to be much lower with a higher recurrence rate in the long term.