

# *Sudden Hearing Loss in the Pediatric Population*

*Kavita Dedhia and David H. Chi*

Sudden hearing loss is rare in the pediatric population. It is defined as a 30 dB loss over more than 3 consecutive frequencies, with rapid onset, ranging from a few hours to a maximum of 3 days. Sudden onset sensorineural hearing loss (SSNHL) is more frequent and better characterized in the adult population. It has a prevalence of 5-20/100,000 adults. It occurs mainly in patients between the ages of 25-60 and peaks at 46-59 years of age. <sup>1</sup> It has been reported that the incidence in adolescents and children is approximately 3.5% of the adult incidence. <sup>2</sup> However other studies still believe the incidence of pediatric SSNHL is largely unknown. <sup>3</sup>, <sup>4</sup> This is hypothesized to be due to difficulty for young children to express hearing loss and the difficulty in performing audiometric evaluation in some infants and young children.

There is a paucity of literature discussing pediatric SSNHL; most of the conclusions regarding management are from the adult literature. The current adult recommendations for management are audiologic testing and magnetic resonance imaging (MRI), to identify brain or retrocochlear pathology. <sup>5</sup> The most significant etiology in adults is due to stroke or retrocochlear lesions. <sup>5,1</sup> In the pediatric population, 20-40% with hearing loss have abnormalities on radiologic testing; however, it has not been determined if computed tomography (CT) or MRI is a better modality for SSNHL. <sup>6,7</sup>

Presentation of sudden hearing loss occurs early in adults, with complaints of hearing loss, tinnitus, vertigo or otalgia. A recent study has shown that there is a delay in presentation in the pediatric population, reporting that 70% presented after 2 weeks of symptom onset. <sup>8</sup> Many factors can contribute to this delay, child not reporting symptoms to caregivers or underestimating patients' complaints. This delay in presentation will delay treatment and can affect outcomes.

Idiopathic viral etiology has also been reported as the most common cause of pediatric SSNHL. However, unlike the adult population retrocochlear lesions and vascular insults are much less likely. <sup>3</sup> Tarshish, *et al.*, identified Lyme disease and Epstein-Barr virus (EBV) as two viral illnesses leading to SSNHL in the pediatric population. They also found that congenital cytomegalovirus (CMV), radiographic findings of enlarged vestibular aqueduct (EVA), and non-organic hearing loss were other causes not often the etiology in adults. Perilymphatic fistula is also a rare etiology associated with SSNHL in the pediatric population. <sup>9, 10, 11</sup> Grundfast *et al* described a series of 5 pediatric patients with perilymphatic fistula as the etiology of SSNHL. <sup>9</sup>

Anatomic abnormalities have also been identified on scans in patients with pediatric SSNHL. Dedhia *et al.*, showed that 40% with abnormal findings on scans. <sup>8</sup> The current American Academy of Otolaryngology Guidelines for adult

SSNHL recommend MRI for SSNHL, as 2.7 to 10.2% of individual with sudden loss are identified to have a retrocochlear lesion on MRI.<sup>5</sup> This is an integral part in the diagnostic testing to rule out acoustic neuromas, which are not common in the pediatric patient.<sup>5</sup> They strongly recommend against CT scans. However in the pediatric population there is not enough data to decide which imaging modality is better. One study found anatomic abnormalities on CT that were not identified on MRI.<sup>8</sup> It may be prudent to perform both in children.

Along with imaging, serial audiometric testing has also been recommended to document progression of hearing loss.<sup>5</sup> There may be a role to perform auditory brainstem response (ABR) in pediatric patients to rule out non-organic hearing loss.<sup>12</sup> Non-organic cause can lead to unnecessary diagnostic testing.<sup>3,12</sup> Studies have shown, routine laboratory evaluation of all patients has low diagnostic yield and should only be performed when there is clinical suspicion.<sup>13,14</sup>

Management of SSNHL is an area of debate. The current available treatment regimens are oral steroids, intratympanic steroids, both oral and intratympanic steroids, and hyperbaric oxygen treatment. Antivirals, thrombolytics, vasodilators, vasoactive substance and antioxidants are not advocated in the treatment of SSNHL.<sup>5</sup> Oral steroids are recommended as the initial treatment.<sup>5,15,16</sup> *Wilson et al* performed a double blind study to evaluate the efficacy of oral steroids. Sixty one percent of their patients improved with oral steroids, whereas only 32% improved in the placebo group.<sup>15</sup> There was also a 10-year retrospective analysis, which found that patients who were treated with steroids showed improvement especially those with severe to profound loss.<sup>16</sup> A study by Chen, *et al.*, is one of the few papers describing the management of pediatric SSNHL in 14 patients with oral steroids given within 14 days of onset.<sup>4</sup>

Intratympanic (IT) steroid injections are an alternative to oral steroids. They provide localized treatment with a higher perilymphatic concentration and avoid systemic side effects. Some of the disadvantages include otalgia during and after treatment, persistent tympanic membrane perforation, vertigo, dysgeusia, and potential infection. Rauch *et al* performed a prospective adult study comparing oral versus intratympanic steroid treatment, over the course of 2 weeks. They showed both treatment regimens to be equally efficacious in the management of SSNHL.<sup>17</sup> It has also been shown to be effective as salvage therapy in patients with severe to profound hearing loss.<sup>18</sup> Using a combination of steroids and IT injections is also discussed in the literature. A study performed by Gundogan, *et al.*, showed more improvement with both oral and IT steroids than either one alone.<sup>19</sup> Only 2 studies have discussed salvage IT injections in the pediatric literature. In one study 2 patients receiving IT injections, one patient with stable hearing after injection and the other with fluctuating loss that improved with subsequent injections.<sup>8</sup> Pitaro *et al* reported 5 out of 8 patients had partial recovery with salvage IT steroids.<sup>20</sup> IT injections in the pediatric patient can be challenging as younger patients may not cooperate with the procedure, compliance issues with frequent visits causing school/work absences.

Hyperbaric oxygen therapy (HBOT) is another treatment modality discussed in the adult literature. This has not been widely used in the United States and is not

approved by the Food and Drug Administration for SSNHL. HBOT reduces hypoxia and edema and may help with immune response to infection and ischemia.<sup>5</sup> There may be a role for use in patients who have failed the traditional treatment regimen.<sup>21</sup> Narozny *et al* found the combination of pharmacologic treatment and HBOT had better outcomes compared to medications alone.<sup>22</sup> The guidelines recommend it as an adjuvant therapy within 3 months in adults.<sup>5</sup> HBOT is expensive, has potentially adverse side effects, and the power of most of the studies is low. Additionally, it may be cumbersome for the pediatric patient.

In conclusion, pediatric SSNHL is a rare entity. It is difficult to report the exact incidence, as very young children may be unable to express hearing loss. This can lead to a delay in diagnosis, which has been identified as a poor prognostic factor. Another unique aspect is the higher incidence of anatomic abnormalities identified on CT and MRI scans. There may be a benefit for obtaining both scans, as some abnormalities may show up only on CT and not MRI and vice versa.<sup>8</sup> This is unlike the adult population, where MRI is the imaging study of choice. Etiology is also different in the pediatric population and is generally not attributed to a retrocochlear or vascular cause. Like adults viral/presumed viral etiology is the most common. Furthermore, pediatric patients also benefit from oral steroids and salvage IT steroid injections. However given the small sample sizes and limited studies on this topic larger studies need to be performed to show benefit of both oral and IT steroids in pediatric patients.

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