Sensorineural hearing loss in sickle cell disease

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ABSTRACT

Sensorineural hearing loss is a common problem in patients with sickle cell disease. The frequency of this problem is ranging from as low as 12% in developed countries and as high as 29% in developing countries. The exact pathogenesis is not known, but it is most likely due to cochlear damage caused by vascular occlusion. Generally, there is no relationship between the frequency of hearing loss and age or sex of the patient, but it may be related to early occurrence of first vaso-occlusive crisis. The onset of this complication is usually gradual and unrecognized. It can be unilateral or bilateral, reversible or irreversible and mild or severe. As sensorineural hearing loss is usually mild and more common to be unilateral, diagnosis may be difficult at the early stage. High index of suspicion among parents, teachers and physicians and annual hearing evaluation may help in early diagnosis. As soon as sensorineural hearing loss is diagnosed, a careful plan of management by both the physician and the otolaryngologist should be put forward.

Keywords: Sickle cell disease, sensorineural hearing loss, hearing loss.


Hearing loss is a common and important worldwide problem. The most consistent and persistent effects of hearing loss in infancy and early childhood are the effects on speech and language development. Hearing loss in the first 3 years of life is associated with lower test scores for speech and language, cognitive ability and school achievement. It also imparts a significant psychosocial debilitation, particularly in young and otherwise healthy individuals.

Most children with permanent hearing impairment suffer from sensorineural hearing loss (SNHL), for which a number of possible causes were reported. One of these causes was sickle cell disease (SCD). SNHL is a recognized complication of SCD since more than 20 years. There is a consensus in the various audiological studies that patients with sickle cell anemia have a much higher incidence of SNHL than the rest of the population.

Sickle cell disease is a common problem in Saudi Arabia, but the reports on the hearing loss from this part of the world, including those due to SNHL, rarely mentioned SCD among these causes. Some of these reports failed to detect the causes of hearing loss in some of their patients, and whether SCD was one of these causes or not can not be verified. This is possibly the same in other countries, where SCD is common.

This subject was not well reviewed and little, if any, was written about it even in those monographs on SCD. We therefore are presenting this review article hoping that it will help the physicians caring for SCD patients in the early diagnosis and management of this important and serious complication.

Definition. Sensorineural hearing loss is a clinical sign which indicates a deficit in the end organ of hearing, the cochlea and/or its neural connections.
Based on the standards set by the American Academy of Ophthalmology and Otolaryngology in 1959, deafness is defined as hearing loss greater than 20dB at two or more frequencies in one or both ears. Patients who had hearing losses involving frequencies above 2000 Hz were classified as high frequency losses.  

**Frequency.** The frequency of sensorineural hearing loss in sickle cell anemia is variable. Studies from developed countries revealed much lower frequency of this problem in comparison to developing countries. Friedman et al reported 12% in the United States of America and Ajulo et al reported 13.5% in the United Kingdom. In developing countries, 21.7% was reported by Todd et al from Jamaica, 21.4% by Odotoyinbo and Adekile from Nigeria and 29% was reported by Atsina and Ankra-Badu from Ghana. Earlier works from Saudi Arabia by Ashor and Al-Awamy revealed 23.8%. The authors have recently reported their experience in Qatif, Saudi Arabia, which revealed a frequency of 19.2%. 

**Pathogenesis.** The pathogenesis of sensorineural hearing loss in patients with sickle cell anemia is not known. A number of hypotheses have been suggested about the pathophysiology of this problem. Koide et al demonstrated experimentally that cochlear venous system had a low oxygen tension conducive to sickling, and ligation of the vena aqueductus cochlea in experimental animals produced changes in cochlear microphonics similar to the pattern of hearing loss in homozygous SCD. 

Morgenstein and Manace examined the temporal bones of a 10 year old boy with SCD and moderate bilateral SNHL who died during a crisis. They found that many of the inner and outer hair cells throughout the organ of corti, were absent or abnormal, cells of the stria vascularis were disrupted and strial vessels were engorged with sickle cells. The bone marrow was hyperplastic and congested with sickle cells. They suggested that the damage to hair cells and stria vascularis was consistent with hypoxic and ischemic damage respectively. They also suggested that expansion of bone marrow in the petrous temporal bone led to narrowing of the internal auditory canal and compression of the eighth cranial nerve. 

Todd et al studied a large sample of Jamaican sickle cell patients for SNHL, and they suggested that hearing loss in these patients is due to continuous low grade venous thrombotic process affecting the cochlea without clinically recognized episodes. 

Berry found that sickle cell patients who experienced more frequent crises showed poorer threshold responses. 

Serjeant et al studied the above mentioned theory of Morgenstein and Manace which stated that expansion of the marrow in the petrous temporal bone led to narrowing of the internal auditory canal and compression of the eight cranial nerve. They found no correlation between abnormal audiograms and narrowing of the internal auditory canals and they concluded that this mechanism is not responsible for hearing loss in sickle cell anemia. On the other hand, they suggested that sickling and impaired blood flow in the cochlear venous system with secondary anoxia of the hair cells and stria vascularis appears to be the most tenable hypothesis for SNHL in homozygous SCD. 

Marcus and Lee reported two sisters with sickle cell thalassemia who complained of vertigo in isolation or vertigo with hearing loss after strenuous exercise. 

Orchik and Dunn reported a total SNHL in an 18 year old male upon recovery from sickle cell crisis, which is similar to the above case reported by Urban. They also reported absent acoustic reflex threshold and zero percent speech discrimination and suggested that this might be due to neural involvement. 

Sharp and Orchik found evidence of unilateral SNHL in one of nine SCD patients. Eight of the nine patients showed normal pure tone sensitivity. On the other hand, 4 patients showed absent acoustic reflex threshold levels as compared to a control group. Also time-compressed speech discrimination was poorer in the SCD patients. These findings can be explained by impaired neural function, as there was no evidence of middle ear involvement. 

Friedman EM et al reported similar findings to Sharp and Orchik (1978) in one of their 5 patients with SNHL who displayed bilateral high frequency hearing loss. They suggested that perhaps slight middle ear involvement not readily apparent from other measurements caused these elevated and absent acoustic reflex threshold, however, these abnormalities also could reflect damage to portions of the cochlea not evident by pure tone air conduction thresholds. 

Odotoyinbo and Adekile reported similar findings to Sharp and Orchik in seven patients, but they failed to demonstrate any recruitment. Five others with similar SNHL who recorded normal acoustic reflex threshold and showed recruitment. They suggested that the damage in these patients is multifocal and involves the cochlea and the retrocochlear structures to varying degrees in different individuals. They also found that 58.3% of those who had SNHL, had their first vaso-occlusive episode occurring before the age of one year and more than 90 percent before the age of five years. They suggested that the cochlear microvasculature in young infants was more susceptible to occlusion during sickle cell crisis and that this manifests as hearing loss later in life. 

Ajulo et al reported also similar findings to Sharp
and Orchik in four out of 12 ears of patients with sickle cell anemia and SNHL. Their conclusions were similar to those made by Friedman et al, and they gave more emphasis to the effect on stapedial function which may not yet be fully understood.

Al-Dabbous et al22 found more than 90% of those who had SNHL, had their first painful crisis before six years of age. They suggested that early exposure of the cochlea to vaso-occlusive episodes may cause cochlear damage, leading to hearing loss. They found also that high level of HbF is protective against SNHL, while associated alpha-thalassemia may increase the risk of this complication.

Some authors (Orchik and Dunn,27 Friedman et al.,17 Odetojinbo and Adekile49) suggested an additional neural involvement to the causes of SNHL in patients with SCD. The consensus, however, is that SNHL in patients with SCD is most likely due to cochlear damage caused by vascular occlusion, although neural contribution can not be excluded. In young patients, the cochlea may be more susceptible to occlusion during sickle cell crises, which may manifest as hearing loss later in life.

Clinical features. Sensorineural hearing loss in SCD may occur at any age and it is as common in males as in females.18,22 The youngest reported patient was 6 years old,19,22 and there is generally no relationship between the frequency of hearing loss and age.6,17,22 There may be a relationship between SNHL and early occurrence of first vasoocclusive crisis.19,22

The onset of this complication is usually gradual and unrecognized,6,17,19,21,22 but sudden onset of hearing loss consequent to sickle cell crises,24,28 strenuous exercise,28 or flying in a modern pressurized jet aircraft49 has been reported.

It can be unilateral or bilateral.6,17,18,21,22,24,28-31 If unilateral, it can affect the right side as often as the left.18,31

SNHL in SCD can be reversible,22,24,31 but it can also progress to permanent hearing loss.22,31 The reversibility of SNHL in patients with SCD is an interesting phenomenon as this may not occur in other diseases.3,32 This was thought to occur because of reversible ischemic changes of the cochlear microvasculature, and it has an important implication in the management of these patients. Reversibility may occur as long as one month after the attack.31 The permanent hearing loss on the other hand may be explained by the cumulative effect of multiple crises.18

The degree of hearing impairment ranges from mild to severe, but usually it is mild to moderate high frequency loss.33 Simple classification6 of hearing impairment is shown in Table 1.

Risk and protective factors. The risk factors of sensorineural hearing loss in patients with SCD are not clear. Review of literatures revealed that certain factors may precede the occurrence of SNHL. The most important factors are shown in Table 2.

There are also a number of possible protective factors against this complication in patients with SCD. The most important factors are good health and high hemoglobin "F" level.6,22

Diagnosis. Sensorineural hearing loss in SCD is usually mild and commonly unilateral. These mild cases may be difficult to diagnose at an early stage. The following points may help to detect early hearing loss in these patients:— 1. Parent’s suspicion: good, educated parents may suspect hearing loss in their child and report early to health professionals. 2. Teacher’s suspicion: Teachers may detect those students with hearing impairment. Poor academic performance, repeating a grade and school behavior problems may be caused by unilateral SNHL.34 3. Physician’s suspicion: A careful physician may suspect hearing impairment if he takes a good history and performs a good physical examination during clinic visit for any complaints. 4. Annual hearing evaluation of all patients with SCD may detect most patients with hearing impairments.

As soon as hearing impairment is suspected, patient should be referred to otolaryngologist for full evaluation. It may be preferable to choose those otolaryngologists who are interested in SCD if they are available.

Management. A careful plan of management by both the physician and the otolaryngologist should be
put forward. Patients with mild hearing impairment may need just reassurance and periodic follow-up. On the other hand, patients with severe hearing impairment may need auditory training, hearing aid, cochlear implantation, speech or occupational rehabilitation. Special education is the mainstay in the management of deaf children. There are several college programs for the deaf. A small minority of deaf students attend regular colleges, but they typically get little out of lecture courses and depend heavily on class notes from hearing classmates, on textbooks and on individual help. Despite poor language and academic skills, the majority of deaf adults are employed and independent.

Periodic evaluation of hearing should be carried out for all patients with any degree of hearing impairment. Reversibility or improvement of hearing would be detected by careful follow-up, and management should be changed accordingly.

A general health care should be provided for these patients. This includes regular follow up preferably in specialized sickle cell clinics. In these clinics education about the disease, ways to prevent sickle cell crises, early detection of complications and early management should be provided. Attention should be made for the psychosocial, educational and financial problems.

A special attention to the mother with SCD or sickle cell trait during pregnancy and at delivery may protect the offspring with sickle cell disease from hypoxic insult and may prevent this complication.

Early administration of the vaccine for Hemophilus influenzae type B, Pneumococcal pneumoniae and Neisseria meningitidis, and oral penicillin prophylaxis may reduce the incidence of meningitis and subsequently reduce the incidence of SNHL in SCD patients.

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References