REVIEW

Some Historical Facts on Otosclerosis in Pregnancy

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Objectieve: To give a historical overview on the different approaches to hearing loss due to otosclerosis in pregnancy in the first half of the 20th century.

Background/Method: A literature study of historical textbooks, thesis and publications concerning the treatment of hearing loss in cases of otosclerosis in pregnancy.

Conclusions: After the early first observations that otosclerosis occurred more often in women and that pregnancy possibly aggravated the hearing loss, treatment of the disease consisted of prevention of conception. Because of the lack of successful surgical treatment, prevention and later influenced by the popularity of eugenics, sterilization and abortion were the options to treat otosclerosis in pregnancy in the first half of the 20th century. The success of surgical treatment by fenestration and later stapes surgery made the relation of otosclerosis and pregnancy less relevant.

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Pregnancy and otosclerosis

Joseph Toynbee[1] from London demonstrated after studying 1149 temporal bones that in 136 cases ankylosis of the stapes footplate could be demonstrated. Adam Politzer[2] performed histological investigations of the stapes and discovered new bone formation of the footplate which received the name otosclerosis. Otosclerosis resulted in bone conduction hearing loss and it was noted that the disease occurs more often in female than in men. Friedrich Bezold[3,4] from Wiesbaden Germany performed a statistical (Fig. 1) analysis in a retrospective study over the period 1884–1892 and calculated the percentage of the female and men affected by otosclerosis. He found that in

Figure 1. Friedrich Bezold (1842-1908)
60.2-61.1 % female and 41.8-58.2 % male were affected. Wolf (1895) observed the influence of pregnancy on the hearing organ. Heimann (1909) stated that the disease occurred twice as much in female than in men. Politzer was one of the first who suggested the relation of otosclerosis to the puerperium and that otosclerosis was a true infective puerperal osteitis. Later Sohier Bryant (1915) mentioned that pregnancy or the puerperium possible intoxicated the temporal bone. The temporal bone might be affected by bony changes which occurred in the puerperium and in that way otosclerosis was produced.

Denker pointed in 1904 on the possible relation between otosclerosis and pregnancy and the puerperium and suggests that in cases of a strong hereditary pathway of familiar otosclerosis the female patient should consider prevention of conception. On one of the meetings of the German otological society he describes the sad side of pregnant otosclerotic patients with a rapid loss of hearing and severe tinnitus. Körner (1914) described in his textbook the observation that the loss of hearing in otosclerosis is often a slow progress but in pregnancy this loss is often fast progressive. He was rather distinct in his thoughts on how to treat young girls who were suffering from hearing loss due to otosclerosis. He advised them not to marry or to get children because every pregnancy could create latent determinants.

Sterilization and artificial abortus in otosclerosis
Artur Blohmke, assistant of the University Ear clinic in Konigberg Germany performed in 1918 an extensive study on pregnancy and the etiology and the treatment in otosclerosis. He did a survey with different German ear clinics with the question whether pregnancy indeed causes a deterioration of hearing in otosclerosis and if in special cases an interruption of the pregnancy was indicated to stop the process of the loss of hearing. Blohmke received many case reports on hearing loss and severe tinnitus due to the pregnancy. In response to the survey it was concluded that there was not an absolute indication for artificial abortus in cases of severe hearing loss and tinnitus in pregnancy with otosclerosis. On the other hand in pregnancy cases of a tendency to suicide because of severe tinnitus and strong hearing loss abortion was sometimes performed. Brickner believed that only interruption of the pregnancy could stop the acute deterioration of hearing problem and that after the abortion the question of sterilization of the female should be considered. According to Blohmke’s survey study Welty performed at least six abortions between 1910 and 1915 because he believed that there was no other treatment to stop the danger of otosclerotic hearing loss due to pregnancy.

Burger from Amsterdam (known of his formula on otosclerosis: “Girls after marriage, Women after pregnancy”) in 1923 described the dilemma’s of a case of a pregnant otosclerotic woman. Although he had not much experience with the abortion treatment for acute hearing loss during pregnancy he suggests after careful considerations and the wish of the patient to do everything to safe her hearing by interruption of her pregnancy. The patient was a musician and her husband had already children of a previous marriage. An abortion was carried out on the 3rd of November 1923. The result of the treatment was no progression of the hearing loss. It seemed that the hearing was improved a bit and that the tinnitus was almost disappeared or only sporadically present. Burger ended in his French publication later with the comment that a brother of the patient visit him later with the same hearing loss due otosclerosis proving the familiar character of the disease in his case. The relation between pregnancy and hearing loss due to otosclerosis as described in the case of Burger was not always that clear. In 1938 Professor Nager from Zurich Switzerland warned against inaccurate declarations in cases of otosclerosis for compulsory sterilization or abortion. He described a case in which a presumably otosclerotic pregnant patient was treated by Röntgen radiation (Röntgenkastrationen) of the gravis uterus according the method advised in the book “Eugenical sterilization in the United States” by Laughlin (1922). This method of abortion had the preference because it was
(Figure 2) believed that procedure resulted in a normal end of the pregnancy. Unfortunately the patient developed a lethal sepsis. Autopsy was performed on the temporal bones and revealed no sign of any histological otosclerosis \(^\text{(13)}\). Nager condemned since compulsory sterilization or abortion in pregnant otosclerosis cases. Nager like Körner supported on the other hand the eugenic ideas of preventive sterilization based on the work of the well-respected Alexander Graham Bell in avoiding marriages and offspring of hearing impaired patients.

**Eugenics and otosclerosis**

Due to his preoccupation with his mother’s deafness Alexander Graham Bell (1847 – 1922), the inventor of the telephone, choose to study acoustics. Because of that interest he investigated the rate of deafness on Martha’s Vineyard, Massachusetts in 1881. He noticed that on that island on the East Coast of the United States a high rate of hereditary deafness existed. The deaf islanders probably originated from immigrants of Weald in the county Kent in England where hereditary deafness was known in the seventeenth century. The islanders even developed their own language with the deaf population. So mixed marriages between deaf and hearing spouses comprised 65% of all deaf marriages on the island in the late 19th century. Compared with the 20 % on the mainland this percentage was far much higher. From this Bell concluded that deafness was hereditary in nature and, that congenitally deaf parents were more likely to produce deaf children. Tentatively he suggested that couples where both were deaf should not marry. At that time deafness was considered something that ought to be eradicated. In his lecture “Memoir upon the formation of a deaf variety of the human race” presented to the National Academy of Sciences on 13 November 1883 Bell defended his thoughts \(^\text{(14)}\). Alexander Graham Bell was one of the earliest modern advocates of eugenics.

Eugenics is the science and the social movement which advocated the use of practices aimed at improving the genetic composition of the human race. The basics of this movement were certain interpretations of the inheritance work of Gregor Johann Mendel (1822 – 1884). Eugenics was widely popular in the early decades of the 20th century. Charles Benedict Davenport (1866–1944) \(^\text{(15)}\), a (Fig. 3)
biologist, was one of leading eugenicists at that time and took eugenics from a scientific idea to a worldwide movement implemented in many countries. After receiving a grant of 500,000 dollar of the heirs of Edward Henry Harriman, a Railroad magnate in 1910 Davenport could build up a research centre called the Eugenics Records Office (ERO) in Cold Spring Harbor USA. Davenport studied the genetics of different diseases including neurofibromatosis, epilepsy, goiter and otosclerosis. He was one of the first to publish a book and an article about the heredity of otosclerosis [16,17]. At the foundation of the ERO Davenport asked Harry Hamilton Laughlin (1880–1943) to become the superintendent and later the director of his research centre for eugenic studies. Alexander Graham Bell was from 1912 until 1918 the chairman of the board of scientific advisers to the ERO. Over 750,000 data were collected and documented about all kinds of genetic diseases.

Davenport and Laughlin organized three International Congresses of Eugenics. One took place in London (1912) and two in New York (1921 & 1932). As a result of all the data collected Laughlin prepared a voluminous book with the solution to racial betterment. His book entitled: “Eugenical sterilization in the United States with a model eugenical sterilization law” was published in 1922 and provided the scientific basis for later eugenic policies such as for example enforced sterilization [18]. The eugenical sterilization law came into force in (Fig. 4)18 states of the USA in 1924. Besides other illnesses and alcoholism also deaf (including those with seriously impaired hearing) were included as subjects for eugenic sterilization. On page 375 in book of Laughlin otosclerosis is indicated as a hereditary disease eligible for eugenic sterilization. (Fig. 5) The influence of the eugenic movement had certainly its impact on hereditary deafness but also on otosclerosis in cases with the combination of pregnancy.

At The Third International Eugenics Congress (1932) in New York presided by Davenport it was emphasized that birth selection better than birth control was the method to better the offspring and to get rid society of “the unfit”. Doom of civilization predicted unless eugenic measures like compulsory sterilization or even abortion were implemented. Laughlin director of the Eugenics Records

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**Figure 4. Cover of the book : “Eugenical sterilization in the United States” by H.H.Laughlin 1922.**

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**Figure 5. Otosclerosis an Eugenical diagnosis indication in the book “Eugenical sterilization in the United States” on page 375.**

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<tr>
<th>DOMINANT</th>
<th>RECESSIVE</th>
<th>REFERENCE</th>
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<tbody>
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<td>Normal condition</td>
<td>Otosclerosis</td>
<td>Lucie, A., 1907, “Die Chronische Progressive Schwerhööigkeit”</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hammerschlag, V., 1905, “Zur Frage der Vererbbarkeit der Otozklrose”</td>
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Office USA made already in 1922 the recommendations for eugenically sterilization. In Germany in 1933 Ernst Rüdin (1874-1952) president of the International Federation of Eugenics Societies wrote for the Nazi’s the (Gesetz zur Verhütung erbkranken Nachwuchses - GesVeN) or “Sterilization Law” which was a modification of the model law by Laughlin[19]. (Fig. 6)

This “Sterilization Law” was a statute in Nazi Germany enacted and was made active in January 1934 by approval of the chancellor of the Reich Adolf Hitler which allowed the compulsory sterilization of any citizen who in the opinion of a “Genetic Health Court” (Erbgesundheitsgericht) suffered from a list of alleged genetic disorders. Any person according this law was considered as hereditarily diseased who is suffering from any one of the following diseases:

1. Congenital Mental Deficiency,
2. Schizophrenia,
3. Manic-Depressive Insanity,
4. Hereditary Epilepsy,
5. Hereditary Chorea (Huntington’s),
6. Hereditary Blindness,
7. Hereditary Deafness,
8. Any severe hereditary deformity.

At pseudo processes of the “Erbgesundheitgerichten” the indication for the sterilization and or abortion was made and so about 400,000 persons were made infertile during the Nazi regime.

At the German scientific otology society meetings at that time compulsory sterilization or even abortion hereditary deafness were often serious topics for discussions[20]. For example Wittmaack (Hamburg) and Kahler (Freiburg) were against this type of treatment of hereditary deafness while Schwartz (Frankfurt) (es bleibt kein anserer weg zur bekämpfung der erblichen taubheit) and Mayer (Wien –Austria) were protagonists of the compulsory sterilization as only solution of eugenic idea.

Greifenstein[21] from Munchen published papers in 1939 about abortion and sterilization in otosclerosis cases as the genetic disease in pregnancy. These papers could be considered to be the guidelines from the German Reichsgutacherstelle to legalize in more detail these

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**Figure 6.** German version of the “Sterilization Law” approved by the chancellor of the Reich Adolf Hitler in 1934.

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treatments for the eugenic reason of terminating the genetic disease otosclerosis. According these guideline forty three pregnant otosclerotic women had an abortion and twenty three were sterilized in 1939. The total number of pregnant otosclerotic woman who were treated by sterilization and or abortion during the Nazi regime will never be known but from the few data available it must have been at least hundreds to a maybe few thousands cases.

In 1938 Guggenheim [22] from Los Angeles (USA) pointed on the primary importance of the avoidance of propagation by otosclerotics. Especially for the otosclerotic women because of two reasons. One reason was that the offspring may be inherit the malady and the other reason was that the mother may suffer a market and rapid deterioration of her hearing as a result of the pregnancy. Guggenheim established a deafness prevention center. The treatment of pregnant women with severe otosclerosis by abortion to stop the hearing loss was in the USA in the forties not an infrequent procedure according to Barton from Boston [23]. He described in 1945 cases in which improvement of perceptive otosclerotic hearing was achieved after so-called therapeutic abortion. In 1951 Pearson [24] wrote a review in which he concluded that because of the otosclerosis and pregnancy often go together, termination of the pregnancy and sometimes sterilization might be considered as treatment in progressive cases of otosclerosis in pregnancy.

Current views on otosclerosis and pregnancy

After the good results of the surgical treatment by stapes mobilization and later stapedectomy the importance of the relation and the role of otosclerosis in pregnancy became less important. In 1954 Walsh [25] already found that there didn’t exists any evidence of a relationship between hearing loss, pregnancy, and otosclerosis. Precechtel (1967) on the other hand found that pregnancy can aggravate the symptoms of otosclerosis [26]. Hall et al (1974) demonstrated that only in 8 % of the otosclerotic patients had an aggravation of hearing problems during their pregnancy [27]. Later in a retrospective study of 479 women with deafness from otosclerosis Gristwood and Venables (1983) demonstrated that pregnancy does involve a risk of aggravating deafness in clinical otosclerosis [28]. They reported that the chance in female patients with bilateral pregnancy-related otosclerosis of subjective deterioration of hearing varied from about 33% after 1 pregnancy to about 63% after 6 pregnancies. In women with unilateral pregnancy otosclerosis this deterioration of hearing was much less commonly observed.

At the introduction of the oral contraceptive pill the fear was raised that due to the hormonal treatment of women increase of otosclerosis could occur. Goethals et al (1963) reported in a pilot study 29% hearing impairment resulted from oral contraceptive pill use [29]. Ten years after this publication Podoshin et al (1974) were able to prove with an extensive survey that the “pill” did not influence the otosclerotic process [30]. In 1984 Kay & Wingrave investigated over 10.000 women and confirmed that the pill did not influence the hearing of women with otosclerosis [31]. More recently Vessey & Painter (2001) also confirmed after a 26 years follow-up of 17.032 women that there exist no negative effect of oral contraception on the hearing function in cases of otosclerosis [32].

Recently Lippy at al (2005) found no adverse effect on hearing in women with otosclerosis who had children compared with otosclerotic women without children [33]. This age-adjusted study of 94 otosclerotic women revealed no negative effect on any outcome measurements like hearing levels and age at time of surgery caused by pregnancy. Also he could demonstrate that there exists no evidence of any deterioration of hearing with single or multiple pregnancies in otosclerotic women.

Final remarks

The historical facts on the struggle to treat hearing loss in pregnant otosclerosis patients are at least interesting. Prevention and later influenced by the popularity of eugenics sterilization and abortion were the options to treat otosclerosis in pregnancy in the first half the 20th century. The success of surgical treatment by fenestration and later stapes surgery made the relation of otosclerosis and pregnancy less
relevant. More recent studies confirm that the role of pregnancy on the otosclerotic process seems to be small and that gynaecological treatments of the past in these cases must now be considered as useless and obsolete.

References
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