Childhood deafness associated with marriage/relationships between first cousins position statement

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1. Purpose

1.1. There has been much debate in the media about the risks of genetic disorders in children of marriages between cousins and the impact on public health and resources[(1)]. The purpose of this statement is to provide information for families, professionals who work with deaf children, and the media, on the risk of deafness in children that may be associated with marriage/relationships between first cousins.

2. Overview

2.1. Deafness and genetics

2.1.1. Just as children inherit features, such as hair or eye colour, from their parents, sometimes deafness is inherited.

2.1.2. In the UK it is estimated that 50% of permanent deafness in children is caused by genetic factors. Sometimes deafness is present at birth and sometimes it happens during early childhood.

2.1.3. Most people have 23 pairs of chromosomes in each of their cells. These are microscopic structures that contain genes. One of each pair of chromosomes is inherited from the mother and the other from the father. Sometimes there is an alteration or change in a particular gene that results in a different characteristic, for example deafness. The change in a gene can be called a 'fault' or 'mutation'.

2.1.4. Genes can work in different ways. Sometimes a child only needs to inherit one faulty gene from either their mother or their father to have that particular characteristic (*dominant inheritance*). Sometimes the child would need to have the same faulty gene passed on from both parents in order to have the condition (*recessive inheritance*). There are other different patterns of genetic inheritance that are less common.

2.1.5. Some people are known as *carriers*. This is where they have the altered gene but do not have the characteristic. So a person who is a carrier for deafness would carry the altered gene, but would not have a hearing loss themselves.

2.1.6. Sometimes a gene mutation is a one-off that simply occurred by chance. This means that, although the hearing loss has a genetic cause, it has happened for the first time in that person and has not been inherited. However, it could be passed onto future generations.

2.1.7. We all carry several altered genes – probably about 3 or 4 on average. For many genes having a second working copy of the gene is enough to make up for an altered copy so that person may never know that they carry it. This is known as recessive inheritance: the altered gene is recessive to the working gene. With recessive inheritance, it is only if someone inherits two altered copies of the same gene, one from each parent, that the effect of the altered copies would be seen. Meeting a partner who carries the same altered gene can happen by chance to any couple in the general population, and often neither parent knows they carry the altered gene until after they have a child with the condition.

2.2. Marriages and relationships between first cousins

2.2.1. First cousins share a grandparent. Second cousins share a great grandparent and third cousins share a great-great grandparent.

2.2.2. Marriages between people who are biologically related as first cousins or second cousins are known as “consanguineous” relationships.

2.2.3. In genetics 'consanguinity' means the amount of shared (identical) DNA. The closer the blood relationship, the more DNA is shared.

2.2.4. For example, second cousins have half the shared DNA as first cousins.

2.2.5. Marriages between first and second cousins account for over 10% of marriages worldwide[9], and worldwide over a billion people live in regions where 20%-50% of marriages are consanguineous[10-11].

2.2.6. In the UK, first-cousin marriages are common among British Pakistanis, East African, Middle-Eastern and Bangladeshi communities. For example, in the UK British Pakistanis constitute 1.5 per cent of the population, of whom about 55% marry a first cousin[9].

2.2.7. Marriage between first cousins is a preferred and longstanding practice between members of some communities who believe that there are significant social, economic and community benefits of this cultural practice. The custom keeps family networks close and ensures assets remain in the family. In Britain, the aim may be to strengthen bonds with the subcontinent as cousins from abroad marry British partners.

2.3. Childhood deafness associated with marriage/relationships between first cousins

2.3.1. Rare recessive genetic conditions and disabilities, such as deafness, are known to appear more frequently in children of first cousin marriages. This is because the parents share DNA and there is a much greater chance of both carrying the same altered gene and therefore passing the condition onto any children they may have.

2.3.2. Recessive genetic deafness affects less than 1 in every 1000 babies born in the general population. Research suggests that children in some ethnic populations in the UK may be between two and four times more likely to be deaf than the general population[2-8] and may affect up to 4 in 1000 babies. So although the risks are higher, they are still considered small. The reasons behind this increase are complex and have not been proved to be solely the result of cousin marriage[2]. For example, it is known that some deafness genes are more common in some communities. This means that families may have higher risk of having a deaf child even if the parents are not related.

2.3.3. There are many other reasons why children are or become deaf including infection, illness, injury, and other types of genetic inheritance. The children of cousin marriages are not at a higher risk of deafness that results from these other causes.

3. NDCS vision and beliefs

3.1. Our vision is a world without barriers for every deaf child.

3.2. We believe:

- Deaf children can do anything other children can do, given early diagnosis and the right support from the start.
- Deaf children should be involved in decisions which affect them at an early an age as possible.
- Families are the most important influence on deaf children and young people, and need clear, balanced information to make informed choices.
- Effective language and communication skills lie at the heart of deaf children and young people's social, emotional and intellectual development.
- Deaf children should be valued by society and have the same opportunities as any other child.

3.3. NDCS's youth strategy also emphasises that deaf children and young people have the right to information and to fully participate in decisions that affect their lives.

3.4. NDCS will continue to campaign for a society in which childhood deafness is not stigmatised and deaf children are viewed as individuals who should have the l same opportunities that are available to hearing people to develop and maximise their skills and abilities.

3.5. NDCS's attitude to genetic developments is based on a belief that deafness should not be regarded as an illness or a life-threatening or life limiting condition. The social model of disability on which NDCS draws on its response to issues relating to genetics, asserts that deafness in itself is not a disability but the way society responds to it does disable people.

3.6. **Informed Choice**

3.6.1. NDCS is committed to empowering parents of deaf children, and deaf children and young people themselves, to make informed choices through the provision of clear and balanced information and support, delivered in a way that is accessible to them.

3.6.2. If asked for advice in relation to first cousin marriages, NDCS will endeavour to ensure that the family receives balanced and clear information about deafness and genetic technologies in order to enable them to make an informed choice.

3.6.3. It is not the role of NDCS to express an opinion on the way families decide to make use of genetic information.

4. Access to genetic services

4.1 The risk of having a deaf child following cousin marriage is a complex and sensitive issue that is best discussed with, and assessed by, specialised genetics services. NDCS believes:

- Families should be offered the chance to discuss their situation with their local clinical genetics (genetic counselling) team.

- Individuals or couples should be offered genetic counselling that is sensitive to their cultural needs, available in their preferred language, including BSL as appropriate, and that enables them to make informed choices for their family.

4.2 *For more information about genetics and childhood deafness, please see Genetic Counselling; Information for families and NDCS Position Statement on Genetics, Stem Cell Therapy and Deafness (2013) You can also call the NDCS Freephone Helpline on 0808 800 8880 or email helpline@ndcs.org.uk.*

4.3 If the issues raised in this paper affect you or your family and you would like specific advice, you should consult a clinical geneticist or genetic counsellor in your local NHS Regional Genetics Centre. To find out where that is, please contact the British Society for Genetic Medicine (www.bsgm.org.uk) on telephone number 0121 627 2634.

5. References

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