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Why tell asymptomatic children of the risk of an adult-onset disease in the family but not test them for it?

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This paper first considers why it is important to give children genetic information about hereditary conditions in the family, which will go on to affect their lives in a salient way. If it is important to inform children that they are at risk for an adult-onset disease that exists in the family, why should they not also grow up knowing whether they actually carry the genetic mutation? Central to this discussion is the importance of the process of disclosure and the environment in which genetic information is divulged. It is concluded that the reasons given for defending disclosure of genetic conditions in the family to children are also important reasons to cautiously defend predictive genetic testing of children for adult-onset diseases.

Genetic service provider guidelines recommend that children should be told that a hereditary disease exists in the family. The Human Genetics Services Association[1] states

> Parents should be encouraged to make their child aware, at an appropriate age, of the genetic condition in the family and the implications, and for the child to be reared with this knowledge. Being able to discuss this information within the family over a number of years at different stages of maturity will ultimately enable the child to make a better informed choice about predictive genetic testing as an adult.

The American Medical Association[2] also claims that if parents discuss the

> … child's risk with the child, they will be able to explain to the child that testing will be available at the discretion of the child when the child reaches maturity.

The guidelines are clear: children ought to be told about genetic conditions that are known to exist in the family, although all agree that children should not be tested for adult-onset diseases until they are old enough to make an informed choice for themselves.

I begin this paper by considering why it is important to give children genetic information that will go on to affect their lives in an important way. Then I ask the question: if it is important to inform children that they are at risk for an adult-onset disease, why should they not also grow up knowing whether they carry the genetic mutation? The debate on whether children ought to be genetically tested for adult-onset diseases is extremely controversial. Many commentators raise major concerns about the child's confidentiality and privacy being violated by such testing, the possibility of genetic discrimination in trying to secure various kinds of insurance and the psychosocial harms that may result from testing. I acknowledge but will not deal with these specific concerns in this discussion as they take me too far from the focus of this paper, which is to critically discuss why it is considered important to disclose genetic information to children and to question whether predictive genetic testing of children is a further step in the disclosure process. It is important to note that many people at risk for late-onset disorders such as Huntington's disease are not aware of their risk status, as the disorder may not have been known about or discussed in the family. For these people, the luxury of choice about whether to be tested is not available.

DISCLOSING INFORMATION TO CHILDREN: WHY TELL THEM?

Why is it important to tell children about genetic conditions known within the family? The Human Genetics Society of Australasia (and others) claims that knowing such information enables the children to make a free choice about genetic testing when they are competent adults. At 18 years old, they are considered to be mature enough to understand what such knowledge means for them and, with support, can come to an informed decision about testing. But we may plausibly suggest that 18-year-olds could be told their risk status and this would still enable them to make a free choice about testing. They would have the rest of their lives to make that choice. Telling older adolescents that they are at risk for an inherited disease may shatter the future they have begun to create and envisage for themselves. By this age, many of them have begun to make definite plans about what kind of career they may follow, whether they will continue with tertiary education or follow a particular career path. Some may have significant partners in their lives and be contemplating starting a family. Others may be more carefree about their future, yet have strong ideas about its direction. Interestingly, it was found that people at risk, who declined to be tested for Huntington's disease, were more likely to have learnt about their risk status during adolescence rather than

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as adults.[3] These people ''suffered severely from the burden of HD [Huntington's disease]'' and were ''significantly more pessimistic about their future''.[3] This may indicate that adolescence may not be an ideal time to disclose important genetic information regarding their risk.[4]

Even though there may be strong reasons to indicate that disclosing certain genetic information to adolescents is not a good idea, it is certainly not clear that disclosing risks of adult-onset disease to young children enables them to make an autonomous choice to be tested later as adults, the child may be told once and it may not be discussed again. Alternatively, the child may not fully understand what such information means. It may only confuse or, worse, frighten the child. Simply telling a child about a genetic condition existing in the family is not enough to satisfy the claim that disclosure of risk facilitates future autonomous decision making as an adult.[5 6]

Although genetic service provider guidelines claim that telling a child about genetic conditions in the family respects the child's future autonomous decision-making capacities (as adults), I suggest there are other important reasons why disclosing genetic information to the child is important.

Secrets and respect

Keeping secrets from children, especially when the secret is about knowledge that will affect the child's life in the future, may result in more harm than good. A secret ''implies intentional concealment of some information important and relevant to the one from whom it is held''.[7] Genetic information is important and relevant to children because it is crucial information about them (as well as other family members) and will possibly go on to have a major effect on their future lives. After all, the fact that something has been kept a secret implies that its revelation will have an influence on the person from whom it has been concealed. Concealment of important information denies children fundamentally important details about themselves and ''long experience demonstrates that hiding information from children usually does not work and that efforts to keep secrets leave children feeling deceived and abandoned''.[8] Skirton[9] recounts the anger expressed by a woman frustrated that her mother persisted in hiding genetic information, which she felt should have been shared. The daughter believed that her mother's insistence on keeping information hidden amounted to her controlling her and her siblings' lives. Parents who know relevant genetic information and who choose not to tell their children who are at risk exert power over them and ''withholding the knowledge from the child also results in a withholding of the power to make informed choices''.[9]

Wright Clayton[8] claims that children are ''not well served by being voiceless until the age of majority'' and that ''keeping information from them is also an affront to their identity''. Telling children about a disease risk in the family may remove some of their uncertainty, especially when they are confronted with extended family members who are exhibiting features of the disease. Children are often aware of tensions in the family without being told any specific information; they may know something is wrong, yet be unsure what it is. Telling children about what is happening in the family, in a way they can understand, respects them as people who have an interest in being informed about information that will go on to influence their future as autonomous agents.

Although there may be differences in disclosing to a young child that he or she has cancer, for instance, and disclosure that a disease is manifest in the family, I suggest they are not relevant differences. Both kinds of information will go on to have profound implications for the child's life, both now and in the future. Children need to be able to make sense of the information given to them, because the disease may have already affected those around them and they need to be able to incorporate such an understanding of what it will mean for their own lives. We simply cannot assume that children are not capable of understanding genetic information or that they are better off not being told when they have experienced the consequences of genetic conditions in their families. Knowing why affected family members react and respond in the way they do may help children understand the family circumstances better.

Claflin and Barbarin, who studied the issue of disclosing information to young children diagnosed with cancer, note that ''lack of disclosure may communicate the unintended message that the disease is a morbid , frightening secret to be worried about and that it is so toxic and dangerous it cannot be discussed openly''.[10]

The Genetic Interest Group response to the UK Clinical Genetics Society, with regard to presymptomatic diagnosis of childhood-onset conditions report, states that

> although the vast majority (of families) would prefer there not to be a genetic disorder in their family, (such) knowledge comes to be accepted as a fact of life in the same way that other issues are recognized to be individual and integral to any family. It is also our experience that children can cope with information about themselves from an early age and that it is much more often the adult who has a problem in giving information.[4]

Biological origins

Disclosure about adoptive and biological origins is considered to be crucial for the child's developing self-esteem and identity. This was recognised in the recent decision of the UK government to reverse anonymity for people who donate germinal material in in vitro fertilisation treatments. Triseliotis[11] states:

> Adoptees who were told or found out (about their adoption) when over the age of about ten felt this deeply and it had a profound adverse effect on them. Revelation at this late stage had a stunning effect, shaking their entire life and self-image, leaving most of them confused and bewildered.

This is also confirmed by Griffith[12]: adoptees who are not told by adolescence ''are much more prone to have greater self-identity conflicts. If not told till after their main self-identity structure is laid down, then they have built the structure on some false foundations. Much pain and turmoil can be caused by late telling''.

Disclosure of disease risk may also benefit children when they are told young as opposed to being told as adolescents or adults. Skirton[9] noted that respondents ''ventured the opinion that it is easier for a young person to deal with the news of risk than it would be at an older age''.

To briefly summarise, children ought to be told about their at-risk status sooner rather than later in their lives. Traditionally, such disclosure has been seen as respecting the child's developing autonomous decision-making abilities. I have claimed, however, that there are other important reasons to think that disclosure of genetic risk is important. If we agree that respect for autonomy is not the only reason to disclose genetic information to children, then perhaps these reasons also apply to predictive genetic testing of children for adult-onset diseases.

Establishing the importance of telling children about genetic conditions in the family is extremely important for the direction and focus of the second part of my discussion, before which I need to highlight and clarify an assumption that cannot be left unchallenged. Disclosure and the subsequent successful incorporation of genetic knowledge into the children's self-identity and the development of their autonomous capabilities assumes that they are told about the disease in a way they can understand. But it is also much more than that. Simply telling children that a disease exists in their family is not sufficient if they are to benefit from such disclosure. Their family and social environment must support the children; thus they need to be surrounded by people who care about them, support them and their family, and are compassionate and sensitive to their needs. The child who is told that Huntington's disease is manifest in the family but who comes to associate this knowledge with fear and uncertainty, the erratic and frightening behaviour of parents or family members, or adults who are distant or who abandon the family when onset begins, will not be empowered by disclosure. A child who is given this information in such an environment may be duly traumatised by such knowledge. Conversely, children who are told that Huntington's disease exists in the family and who see extended family members caring for one another, where support, both practical and moral, is freely given, where the disease is considered simply to be a part of their life and the family is accepting and not fearful of it, is I believe more likely to incorporate such knowledge positively into their self-identity.

IS PREDICTIVE GENETIC TESTING THE NEXT STEP?

An obvious question then arises: if disclosure of disease risk is important because it enables the children to make a free choice about whether or not to be tested as adults, and helps the children to successfully incorporate such knowledge into their self-identity, why not test at-risk children for their genetic status? Although it is true that they will not be able to make a choice not to be tested as an adult, knowing their genetic makeup may benefit them in two major ways: firstly, by offering them important choices that would otherwise be denied to them if they did not have such (genetic) information and, secondly, that they would have grown up in an environment having always known and thus would have assimilated such knowledge into their self identity: the knowledge simply becomes a part of who they are.

Many children who are at risk for an inherited disease will not in fact carry the genetic mutation and so are free of ever developing the disease as an adult. If tested and found not to carry the mutation, they will grow up being able to make different choices—for instance, about reproduction—from those they may have made were they uncertain of their genetic status. As Robertson and Savulescu note, the child who knows the ''truth about herself'' will come to have different rather than fewer choices. These include choices about career, financial planning and end of life decision making.[5 13] Furthermore, the parents of children who do not carry the genetic mutation presumably will not constantly look for signs, which, before testing, they may have interpreted as symptoms of the disease.

Is not testing at-risk children for conditions known to exist in the family simply an extension of the disclosure process? Before answering this question, we must make it quite explicit what manner of disclosure is at issue. Although the guidelines recommend disclosing a disease risk to children, they do not detail how children are to be told, and when or what kinds of information should be given to them. But it follows from concerns about informing children that disclosure must take a fairly ordered and planned path. As I have already mentioned, it is not sufficient to tell children once that a disease is manifest in the family and then leave them to find out on their own what the risk may mean for them. This will not nurture or facilitate their autonomous decision-making capabilities. Disclosure surely requires that the children understand what they are being told, can make sense of the information according to their age, are not fearful of it and are able to talk about it and ask questions whenever they like. If this is what is intended by the process of disclosure, then in such a supportive and enabling environment, is testing a natural next step? For if the children are told of their risk status in a supportive and caring environment, getting them tested may be understood to be a part of the disclosure process. Presumably, the guidelines would advise parents to seek professional help on how to disclose such information to their children and extended family members. Families that are dysfunctional and have trouble discussing personal information that affects their members will no doubt have difficulty disclosing genetic information to young children in an empowering way. Adults who are pessimistic about their own genetic risk, who feel burdened by the disease and who do not envisage a positive future for themselves or their children, may not communicate such risk positively or effectively to their children, if at all. Thus, the testing of children for their risk status would not be advisable in such situations, as it is not in the best interests of the children. It is also doubtful that disclosing a disease risk to children in such an environment is appropriate or in the best interests of the children.

We may plausibly object that there is considerable difference between knowing that you are at risk for a particular disease and knowing for certain that you have the gene that will one day develop into the disease. If you grow up knowing the risk, you can hope that you do not carry the mutation. Being told that they are at risk but not being tested gives the children a certain amount of knowledge, while retaining a degree of hope that they will not harbour the mutation. We may reasonably claim that being tested and knowing for sure that the children have the mutation may foreclose any hope by presenting the children with a doomed future in which their genetic constitution may mean disability or premature senility and death.

I will make two comments in response to this objection. Firstly, it does not follow from the first claim (knowing only that you are at risk) that genetic certainty (you carry the mutation) forecloses any further hope or indeed escalates your sense of hopelessness. Knowing your genetic inheritance may resolve uncertainty and allow the children to plan their life, still hopeful that a cure may be found, that medical science may find a way of halting the progression of the disease or alleviating the symptoms that appear around onset. Hope does not exist solely in the desire not to be afflicted with the gene; it comes in many forms: ''hope that I will not develop symptoms until I am much older'', ''hope that my symptoms will be mild'' or ''if I keep fit and healthy, I hope I may be able to delay the onset''.

Secondly, and related to the first comment, although it is difficult to extrapolate the conclusions reached from studies on adults to children (who are at risk for genetic conditions), we can make some useful observations. Meiser and Dunn,[14] in their review of the literature on Huntington's disease, found that ''although risk factors for psychological sequelae have been identified, few adverse events have been described and no obvious contraindications for testing people at risk have been identified''. This has also been confirmed by others.[15–22] The concern that people who are positive for the Huntington's disease gene face an increased risk for suicide has ''proved to be a rare event''.[23] Of course, it must be remembered that the adults who choose to undergo testing

have self-selected and presumably feel confident enough in their ability to cope with the outcome, be it a negative or a positive test result. However, evidence in the literature on Huntington's disease suggests that those who undergo genetic testing do not lose hope; knowing that they carry the gene for the disease delivers many benefits. Although many late-onset diseases cannot be treated or their progression halted, knowing that they have the genetic mutation may allow people greater control over the rest of their life's plans. For instance, although people know that they are likely to develop a disease in a certain number of years, such knowledge may give them greater impetus to achieve personal success in other aspects of their life (eg, travel and education).

CONCLUSION

International genetic service provider guidelines claim that disclosure of disease risk enables children to grow up being able to make a free choice about genetic testing when they are competent adults. What is being claimed here is that children reared with such knowledge can develop the autonomous capability to make informed decisions (as adults) by being supported and encouraged to understand what such knowledge may mean for them.

However, I have argued that such a process of disclosure may not necessarily exclude predictive genetic testing of children for adult-onset diseases. Being reared with such knowledge may give the children time to adjust to the information and provide them with a range of important choices they otherwise may not have. Rather than having to confront whether or not to be tested as an adult and face the uncertainty and fear that may accompany such a choice—especially when they may have put life plans in place—already knowing their genetic inheritance gives the children different important choices to make as adults. In some families, testing children for adult-onset diseases may be seen as an extension of the disclosure process and thus is in the best interests of the children. The reasons given for defending disclosure to children of genetic conditions in the family are also important reasons for cautiously defending predictive genetic testing of children for adult-onset diseases.

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