

FH podcast #3 FH and Children Transcript
American Heart Association & The FH Foundation

Recording: 00:02 Welcome. You're listening to a series of four familial hypocholesterolemia podcasts, brought to you by the American Heart Association, and the FH Foundation. This series is focused on educating patients, caregivers, and health care providers on ways to improve awareness, detection of FH, and management of high cholesterol.

Katherine Wilemon: 00:23 Hi everyone. My name is Katherine Wilemon, I'm the founder and CEO of the FH Foundation. I also have familial hypocholesterolemia. Today, in our third podcast, it'll be all about children and familial hypocholesterolemia. Joining me is Dr. Samuel Gidding, who is a pediatric cardiologist and the Chief Medical Officer of the FH Foundation. Thank you Dr. Gidding for joining us for this discussion on children and FH.

Dr. S. Gidding: 00:54 It's a pleasure to be here, Katherine.

Katherine Wilemon: 00:58 Before we get started, could you briefly tell our listeners what familial hypocholesterolemia is?

Dr. S. Gidding: 01:07 Familial hypocholesterolemia is a genetic condition where the gene for FH causes very high cholesterol. Cholesterol that is so high and it's not related to diet or being overweight. Familial hypocholesterolemia is present in about one and 200 to 250 individuals in the United States, and it's underdiagnosed, was underdiagnosed and undertreated.

Katherine Wilemon: 01:37 One in 250 people, that's quite a bit. Is it considered a rare condition?

Dr. S. Gidding: 01:46 Historically, it's been considered rare because it was thought to be present in about one and 500. But now, it's the most common genetic condition that causes early death. The early death is from a heart attack, which can occur in middle age or even younger, in people who are effected. The problem with FH is that there's no symptoms. As opposed to other genetic conditions, like marfan or sickle cell, or cystic fibrosis. They're not easily recognizable, FH is not easily recognizable based on symptoms.

Katherine Wilemon: 02:25 Are there other causes, Dr. Gidding, for high cholesterol in children?

Dr. S. Gidding: 02:33 Genetic causes at the level of FH are far and away the most common. The second most common is diet. People always associate obesity with high cholesterol, but in fact, obesity does not cause as severe elevation in cholesterol that FH does. The

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lipid problems in FH relate to other fats in the blood, as well as cholesterol.

- Katherine Wilemon: 02:58 When we're talking about these high levels of cholesterol in children that don't have anything to do with being overweight, what kinds of levels are we talking about? What does high cholesterol in children look like?
- Dr. S. Gidding: 03:11 Typically, the cholesterol, the LDL cholesterol, the bad cholesterol would be over 190, where total cholesterol over 250. To put this in perspective, the average cholesterol in a child in the United States is about 90 to 100, so it's about twice as high. Children with familial hypocholesterolemia also can have cholesterol levels over 160. The current guidelines for pediatricians, is that a high cholesterol level is over 130 for LDL, or 200 for cholesterol. The current guidelines will capture children in the United States with familial hypocholesterolemia.
- Katherine Wilemon: 03:52 I have two children and one of them has FH, but I actually had to ask for my children to have their cholesterol levels tested. As a pediatric cardiologist, do you think that most children are being screened as the guidelines recommend for high cholesterol?
- Dr. S. Gidding: 04:15 The universal screening guideline was published in 2011, and at that point less than 10% of pediatricians were screening all children at age nine to 11. Now, the uptake of that has improved, and we believe that the percent of children who are having their cholesterol tested between nine and 11 is increasing a lot. It's probably still less than half. It's important to know, however, that in your situation, where you have genetic high cholesterol, pediatricians should be checking your children over the age of two years. If it's known that a family has early heart attacks, or very high cholesterol, the cholesterol can be checked anytime after the age of two years.
- Katherine Wilemon: 04:59 FH is a genetic disorder where individuals, including children, have very high cholesterol. What do parents with FH, like myself, need to know about how to get their children screened?
- Dr. S. Gidding: 05:17 Sure. I think that the most important and first thing that parents need to know, is that recognizing FH is important because early treatment prevents heart attacks. We now know that if you begin treatment in teenage years, or over the age of 10 or eight, which is recommended, that your likelihood of having one of these early heart attacks goes down precipitously, as much as 75%. Parents should want the cholesterol in their children

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tested to look for FH. Hopefully, the pediatrician will be doing it at the recommended age, which is nine to 11. However, just as you did, parents who know they have FH should insist that their children have their cholesterol checked.

Katherine Wilemon: 06:09 You and I know that FH is actually highly under diagnosed, but what should make families suspect that they might have FH and need to have themselves and their children screened?

Dr. S. Gidding: 06:23 The most important things for parents, are knowing their own cholesterol and knowing their family history of heart disease. If there's early heart attacks in a lot of family member, or your cholesterol is in the FH range, you should definitely have your child tested. Those would be the most important things.

Katherine Wilemon: 06:44 It's always a serious consideration if you find out that your child does in fact have a disorder, and I can tell you as a parent, it's even hard to realize that you might be the one who has given that to them. But there's also another tough decision, parents don't want to give medicine to their children that isn't absolutely necessary. We hear from many families at the FH Foundation that they struggle with what to do if their child has FH. When to treat them, what medications are appropriate. Could you share with us the safety and reasons for treatment? What do we know about treating FH in children?

Dr. S. Gidding: 07:29 Sure. I think starting at the beginning, any parent who has FH has a 50% of their child having FH. When a parent goes to the doctor about half will be effected, and half will not. It's always very reassuring to find your child does not have high cholesterol. Once the high cholesterol is identified, we recommend beginning treatment at age eight to 10, and the reason for that, is that's about the age when you can tell that the early atherosclerosis is beginning to build up. There's studies which compare the blood vessel thickness of children with FH with their brothers and sisters, and those studies show that beginning at about age 10 or so, that's when you can see a difference in the thickness of the blood vessels. That's why we recommend beginning treatment about age 10.

Dr. S. Gidding: 08:21 With statin treatment, which is far and away the most effective, we know that we can essentially bring the cholesterol levels down to the high normal range for United States children. Parents do have a lot of fears about life long medication, but the realities are for statin use in children, that the doses are much lower than you would use in adults. That's one important fact, and the second thing is, there have been many studies and

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randomized trials of statins in children and what's been observed is actually the side effect profiles are much lower than the side effect profiles in adults, particularly those over age 55 or 60 years.

- Katherine Wilemon: 09:07 There is data from other countries showing that, in fact, it is safe to give children with this genetic condition statins from an early age, you said eight to 10 years old? And that it does reduce significantly their chances of having an early heart attack or very aggressive heart disease?
- Dr. S. Gidding: 09:32 Sure. Now we have data from over 20 years of follow up in children who've started on statins in adolescents, back in the 90s. There's now emerging data from large United States registries and other case series, that there's very little toxicity of statins. In fact, it's unclear if a case of diabetes has ever been reported, for example, in a child on statins and they don't have the muscle pain or other side effects typically experienced by adults.
- Katherine Wilemon: 10:07 What kind of doctor, if someone's child has FH, should they be seeing to manage this genetic condition?
- Dr. S. Gidding: 10:16 Most importantly it's a doctor who has an interest in cholesterol or blood lipids. In the United States, right now, cardiologists, or some cardiologists with reasonable experience, some endocrinologists, some gastroenterologists, and there are some primary care physicians, actually, who have taken an interest in this disorder. I think the most important thing to consider is whether your doctor has experience managing FH, understands the condition and knows the treatment recommendations.
- Katherine Wilemon: 10:50 The FH Foundation has an FH specialist map, and it is our number one resource that people come to because, as you said, it's not a single specialty of physicians, but actually there can be a variety of types of physicians who are well versed in FH. The same is true for treating children.
- Dr. S. Gidding: 11:14 I'm the leader of a very large, or a co-leader of a very large group of pediatricians who are lipid specialists across the country. I think we have over 90 people across the United States who are very interested in the care of familial hypocholesterolemia.
- Katherine Wilemon: 11:32 How do you explain this to a child? Unlike Type I Diabetes, FH is about as common as Type I Diabetes, but there aren't symptoms, and to get a child to start taking medication and to

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take it every day for the remainder of their lives, how have you done that successfully as a pediatric cardiologist?

Dr. S. Gidding: 11:54 I think the important thing to remember about FH is it's really a familial condition. Most of the time within the family, many people are actually wrestling with FH, taking statins, and things like that. I think the first step is to bring the child's experience into the context of the family's experience. One of the things we found particularly in families where there's been a lot of early heart attacks, is the parents actually feel safer when their child is taking the medication. Also, explaining how atherosclerosis develops and why it's asymptomatic is important. Another key thing that I like to emphasize, is that really the genetics create a condition of very high LDL cholesterol, and that's a risk factor. It's the heart attack that's a disease, and if you don't want to have a heart attack, starting treatment very early in life, will give you life long lower cholesterol and dramatically reduce the likelihood of having a heart attack.

Dr. S. Gidding: 12:56 These are the key explaining factors, sometimes we do it over several visits, so I don't necessarily start medication the first time I see the child, but give the family time to talk about it. Genetics counselors often can provide useful information and we also have psychological resources. Really the purpose of the psychological resources isn't so much to get the kid to take a medicine, to understand the full family and many of the issues that could be going on within the family, with regard to an inherited condition dealing with heart disease, dealing with financial issues. There's a pretty broad range of family bases issues that need to be covered in FH. Unfortunately, in the US Healthcare system, our care is very fractured and it's very hard to provide these engraved family resources. I think that's the biggest problem.

Katherine Wilemon: 13:59 One of the things that we do at the FH Foundation is actually connect families who might be in a similar place, and we have found that just like the American Heart Association, I'm sure does, that community also plays an important role and if you're living with a life long condition, you're a different patient at different times. I think having the entire family involved, and the larger community, can be very supportive.

Dr. S. Gidding: 14:33 Another thing is to deliver a positive message so that, we're at an interesting place in history with regard to familial hypocholesterolemia. Right now, for adults, having the FH gene triples your risk of having a heart attack at any level of cholesterol. However, we now have a tremendous opportunity

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through Cascade screening and genetic screening, to identify everyone in the US, or most everyone with FH, and if those individuals start on early treatment, people with FH may end up having less heart attacks than the rest of the population. We're in, I think a 20 to 30 year window of opportunity to go forward.

Dr. S. Gidding: 15:15

Another key thing that I've realized that we haven't discussed, and what's very common in my practice, is a pediatrician will screen a child with high cholesterol, find out they might have FH, we in fact diagnose FH and then test the parents and find out that there's a parent with FH as well, who's not receiving treatment. Another key advantage of identifying children is if their parents haven't been checked to find those parents who are also under treated. In fact, its been shown that for about every child you find with FH, you find one parent whose not treated. Sometimes screening a child for FH actually identifies two people, not one.

Katherine Wilemon: 15:59

That's what we always say. You never find an individual with FH, you always find a family. It truly is a family condition, and as you said, I think that's one of the things also, its been a huge burden to these families, but can be a blessing that you can help an entire family change their health and their future, if they're identified and managed early. It's genetic, it requires medication, but what about diet and lifestyle?

Dr. S. Gidding: 16:31

Diet is actually really important. Though diet can't lower your cholesterol as much as a statin, it can have a big impact. It can lower your cholesterol as much as 10%, which is a 25% risk reduction. There's been some recent genetic studies, which have looked at the relationship of diet and changes in lipids to the presence of genes that raise your cholesterol and what's been shown, is there's actually a bigger impact of diet if you have a high cholesterol. There's this gene environment interaction. Another thing to remember, is that risk is relative. If you have FH, you have a very high risk of having a heart attack, and if you drop your risk by 25%, that actually is a huge reduction.

Dr. S. Gidding: 17:20

If you have low cholesterol and have very low risk of having a heart attack, doing a good diet and lowering your risk by 25%, doesn't have nearly as big an impact on heart attack rates. Another key thing to remember is the mechanism of lowering cholesterol through diet, actually helps statins work because it takes away another source of cholesterol from the liver, and the liver will then be forced to reach out and draw more cholesterol from the blood, and that's the way statins work, is taking the

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livers residual ability to get cholesterol from the blood and putting it into overdrive. Diet and statins are actually more effective together than separately.

- Katherine Wilemon: 18:08 How does the future look for children living with FH?
- Dr. S. Gidding: 18:14 I think the future can be extremely bright. I think that essentially high LDL cholesterol, which you see in FH, is a risk factor. That risk factor can be obliterated, you can have a normal life expectancy. When I started in this field about 30 or 35 years ago, I thought that if I could my prevention patients to age 60, it was somebody's problem. Meeting people with FH, I don't think they ever want to have a heart attack, and I think that's the goal we ought to be looking towards, and I believe it's achievable.
- Katherine Wilemon: 18:53 The first thing we have to do is find them though, right? The biggest barrier to-
- Dr. S. Gidding: 18:56 That's for sure. That's for sure. If you have it, and you don't know you have it, you're behind the eight ball.
- Katherine Wilemon: 19:02 Right. And that's why screening is so important, and like you said, hopefully if you find either a parent or a child first, you can find the rest of the family and help them. Well, I can't thank you enough for all the amazing information that you've been able to share with us today, Dr. Gidding. Thank you. In case you missed any of what we discussed on this podcast today, please visit the FH Foundation at thefhfoundation.org, and the American Heart Association at heart.org/cholesterol. Thanks again for listening, and keep your ears open for the fourth podcast in this series about managing familial hypocholesterolemia.
- Recording: 19:48 Views expressed in this podcast do not necessarily reflect the official policy or position of the American Heart Association, and American Stroke Association. For transcripts of this podcast and more information about cholesterol, please visit heart.org/cholesterol, or engage with us via social media using the hashtag [#understandcholesterol](https://twitter.com/understandcholesterol).