WHAT CAUSES CHILDHOOD STROKE?

Pediatric and adolescent stroke is different from adult stroke in frequency, presentation, management, and prognosis. What causes childhood stroke? How can we diagnose and treat the condition and how do our young stroke patients fare as they get older?

DR. JENNIFER SHU:
Welcome, Dr. Roach. Dr. Roach, you are the lead author of A New American Heart Association Scientific Statement on managing stroke in infants and children. How does childhood stroke differ from adult stroke?

DR. E. STEVE ROACH:
Probably the biggest difference that comes to mind is in the underlying causes that lead to the stroke to begin with. We almost never see atherosclerosis in children as a cause of stroke. We don't see atrial fibrillation very often as the cause of stroke. Those are arguably the two most common underlying factors for ischemic stroke in adults. Children tend to have sickle cell disease, congenital heart disease, other things altogether, that's probably the biggest single difference.
DR. JENNIFER SHU:
Now, the World Health Organization defined stroke as a clinical syndrome of rapidly developing focal or global disturbance of brain function lasting longer than 24 hours or leading to death with no obvious nonvascular cause. Now would that be the same for children as well?

DR. E. STEVE ROACH:
Pretty much. Children's brain has to have blood supply as well and the definitions are about the same. Sometimes, it's a little harder in children to know exactly what you are dealing with in terms of the length of the symptoms and things of that sort, but the main difference there is just in the way we apply the definition or the way we struggle to know what the parameters are, but the definitions are going to be about the same.

DR. JENNIFER SHU:
How common is the childhood stroke in the US?

DR. E. STEVE ROACH:
The number keeps shifting about a little bit. Part of the problem is in years past, people have failed to recognize it and therefore it doesn't get recorded and the numbers tend to be inappropriately low. We would estimate just in rough terms that all forms of childhood stroke including children, neonates, hemorrhagic stroke, ischemic stroke, so on would be approximately twice the rate of brain tumor.

DR. JENNIFER SHU:
Okay, and one statistic I saw is that it's about 2 to 3 children per 100,000 per year, does that sound about right?

DR. E. STEVE ROACH:
That's a relatively older number, but yes that's in the literature, there are other numbers where probably more children were included. Actually, I would suggest a rate of about 2 to 3 times of that actually, but that 2 to 3 is an older number. As I would say it's a little bit of a hard number to get your hands around.
DR. JENNIFER SHU:
The physicians don't need to be on the lookout for it for sure?

DR. E. STEVE ROACH:
Sure.

DR. JENNIFER SHU:
Now, who tends to get childhood stroke, are there certain groups that are at higher risk?

DR. E. STEVE ROACH:
Yes, the risk tends to parallel the risk factors and so children with sickle cell anemia have extraordinarily high rate. I have seen estimates that over the course of the life term with a predilection toward earlier years that as many as 20% to 25% of the children with sickle cell anemia would have a stroke. Children with congenital heart disease, particularly the cyanotic forms, have a very high risk of stroke, although that's reduced considerably if the heart lesion can be fixed and then children with congenital anomalies with the blood vessels, arteriovenous malformations, things of that sort have a very high risk of hemorrhagic stroke. So, basically the people who are at greatest risk are the people who have the major risk factors

DR. JENNIFER SHU:
Now, is that more common in boys or girls and what about younger versus older children?

DR. E. STEVE ROACH:
For the most part, it's not anymore common in boys than girls. I mean certainly nothing that's striking. It's more common in younger children in as much as ischemic stroke is so much more common in neonates than it is in older kids. Once you get out of the neonatal period, it's probably not that much difference say between a 6-year-old and a 12-year-old.

DR. JENNIFER SHU:
Does family history of stroke play any role?
DR. E. STEVE ROACH:

In some instances, but generally no. I mean if a child's grandfather had a stroke when they were 50 that basically carries almost no weight. If a child's sibling had sickle cell disease and they have it too, obviously that does you know, and there may be some slight trend you know based on coagulation anomalies, things like that, but it's not that major a factor. We are often asked say when a child has a stroke in the family or ask very directly, what does this mean for my other children and we usually are pretty reassuring because unless there is some known risk factor that is shared that really increases the odds of that second child within that family having a stroke is pretty low really.

DR. JENNIFER SHU:

Earlier you mentioned that some stroke may be going unrecognized. How do children present with a stroke? Is it pretty obvious or can it be tricky?

DR. E. STEVE ROACH:

It can be tricky. It's always amazed me though. I have studied stroke for 20 years, and if you take in the same family someone's elderly grandparent and if they suddenly lose the ability to use the right side and become unable to speak, everybody in the room says, "quick, let's get grandma to the doctor because it looks like a stroke." You let the same exact phenomenon happen in a 10-year-old in that same family and there is much less urgency. It's like well see what's going on and they might go to the doctor later, but not necessarily as quickly. So, clearly there is a delay in seeking medical attention that's been demonstrated you know formally and published among children versus adults. To some extent, there is still some unwillingness to consider this diagnosis among physicians, although I think that's actually changed a great deal, improved a great deal in the last 8 or 10 years, but children like adults present mostly with focal neurologic deficits. Sometimes the children, the older ones, as well as the younger ones will present with epileptic seizures after a stroke, but for the most part they will have some change in function, some loss of function. The most common of which is weakness on one side of the body. Other things would include double vision, clumsiness, and alteration in consciousness. You know the list of things that could represent a stroke are pretty long because it really reflects where is the stroke going to be and what is the function of that part of the brain that gets knocked out, but the most common is hemiparesis.

If you have just joined us, you are listening to The Clinician's Roundtable on ReachMD. I am your host, Dr. Jennifer Shu. Our guest is Dr. E. Steve Roach, Professor of Pediatrics and Neurology at the Ohio State University College of Medicine and Chief of Neurology at Nationwide Children's Hospital in Columbus, Ohio. We are discussing childhood stroke.

DR. JENNIFER SHU:

Now you mentioned some of the common risk factors being sickle cell disease or congenital heart disease. If these are medically
managed, is it possible to prevent stroke in these children?

DR. E. STEVE ROACH:
Absolutely and it depends on the degree to which you can manage the complication. The best information is actually available for the children with sickle cell disease and the overall risk they are untreated is probably about 25% over the course of lifetime. However, a child who has had stroke #1 because of sickle cell disease has arguably a 50% to 65% chance of having additional strokes, and for many years, we have used blood transfusions to lower the percentage of sickle hemoglobin and that really reduces the stroke risk. It doesn't eliminate it absolutely, but it probably cuts the second stroke risk from 50% to 65% down to 5% or less and that's actually been very thoroughly studied in a controlled clinical trial that was published about 10 years ago, and actually, if you can identify the children who are at high risk for a stroke even before the stroke, then beginning transfusions for those kids will actually prevent the first stroke.

DR. JENNIFER SHU:
Is there anything you might expect to see on physical exam that might predict a stroke risk?

DR. E. STEVE ROACH:
Basically on physical exam, you would be looking for evidence of stroke risk factor. If you have, for example, evidence of congenital heart disease, you can predict that that child is at least going to be at increased risk for stroke. I am sure there are probably some others part of it.

DR. JENNIFER SHU:
Now, The American Heart Association Statement mentions Moyamoya disease, which we all learned about in medical school, but can a physician in the US really expect to see that or is it pretty rare.

DR. E. STEVE ROACH:
A physician who is in practice will see it whether they recognize it or not is another question, but it's not so rare that it's not going to occur. It's much, much more common in individuals of age inherited. There is an increased risk in children who have Down syndrome. There is an increased risk in children who have neurofibromatosis; having received cranial therapeutic radiation sometimes will trigger this. So, you do see it in this country and it's probably not as unusual as we once thought.
DR. JENNIFER SHU:
So, if you see a patient with Down syndrome, for example, should we be doing some type of diagnostic exam to see if they might have Moyamoya disease?

DR. E. STEVE ROACH:
We don't currently and the reason we don't is because even though it's generally agreed that the frequency of Moyamoya syndrome is greater in children with Down syndrome, it's still not extraordinarily high and it may be as high as 3% or 4% even, but that's still pretty low number as the percentage of all children with Down and then you also have to factor in that even doing an MRI and MRA in a cognitively impaired child is going to be a little bit of a big deal sometimes. So, typically what we do is we start investigating after there has been some evidence of a neurologic deficit.

DR. JENNIFER SHU:
So, in any child who is suspected of having stroke, what would be the first diagnostic step to confirm that?

DR. E. STEVE ROACH:
Usually for practical purposes, it's a CT scan and the reason for that is most of these kids even though they don't do it quite as promptly as I would like will eventually show up in an emergency room, and in my experience, the emergency room staff are going to do a CT scan in a child with a new neurologic deficit and they often do that even before they call a neurologist. So, most of these kids are going to get a CT scan first and MRI scan is most of the time more helpful, although it's often not available in the middle of the night and it takes a bit longer. So, the sedation is a little more of an issue, so usually you will get a CT and an MRI in most of these kids. Then depending on the degree to which you are concerned about individual blood vessels, then those kids are going to go on and get like an MRA or CT angiogram or even an old-fashioned catheter angiogram.

DR. JENNIFER SHU:
Now, what would be the acute management recommended for children with stroke?

DR. E. STEVE ROACH:
Depends on the type of stroke than it depends on underlying cause, which in turn is going to drive your concern about additional strokes. The stroke itself realistically there is not a whole lot you can do about that, I mean supportive management, oxygen, make
sure they don't get massive cerebral edema and hernia, but when the therapy is directed toward most of the time is eliminating the risk of an additional stroke and that is almost certainly going to hinge on what you think caused the first stroke. So, for example, a child who comes in and either is known already or subsequently determined to have congenital heart disease, a good number of those kids will get anticoagulated in contrast someone who comes in who had hemorrhagic stroke, none of them are going to get anticoagulated. So, it kind of depends.

DR. JENNIFER SHU:

I would like to thank our guest, Dr. E. Steve Roach. We have been discussing childhood stroke. I am Dr. Jennifer Shu.

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