BroadcastMed | Vision Loss Caused by Retinitis Pigmentosa Improved With Retinal Prosthesis

DR. RAYMOND IEZZI: Hello, I'm Dr. Raymond Iezzi from the Mayo Clinic here in Rochester, Minnesota. I'm vitreo retinal surgeon in the Department of Ophthalmology. And I'd like to talk to you today about a service that we provide to some of our patients with the most severe forms of vision loss due to retinitis pigmentosa.

As you may know, retinitis pigmentosa is an inherited retinal generation that affects 1 in 4,000 people worldwide. A subset of those patients, however, may go on to lose all of their sight. Fortunately, this is the exception, and not the rule. But until relatively recently, we have not had a means of caring for these patients in a manner that would restore their sight.

It's been our dream for several decades, and perhaps hundreds or thousands of years, to be able to restore the sight to the blind. And fortunately, we have access to a retinal prosthetic device that allows us to restore rudimentary forms of vision to those who have lost their sight due to advanced retinitis pigmentosa.

Patients with retinitis pigmentosa fall into several categories. Certainly, there are those with dominantly inherited mutations. And there are those that have recessive mutations. Typically our patients with dominantly inherited disease often retain their sight for most of their life, if not all of their life.

However, our patients with recessively inherited retinitis pigmentosa may go on to lose their sight, even at a young age. And it's probably that population of patients who most likely would benefit from a retinal prosthetic device. Many, however, often develop retinitis pigmentosa as a simplex condition, a spontaneous mutation. And we may not identify a specific pattern of familial inheritance. And some of those patients may also go on to require a retinal prosthesis and others not.

So how is it that a retinal prosthesis works and who might benefit? Well, we know that retinitis pigmentosa involves a degeneration in the photoreceptor layer of the retina. This is the outermost layer of the retina, adjacent to the retina pigment epithelium. And it is the portion of the retina that undergoes the most rapid degeneration.

Scientific studies have found that the inner retina, the ganglion cell layer, the layer that forms the optic nerve and the connection of the eye to the brain, the optic nerve layer remains relatively intact in almost everyone with rhinitis pigmentosa. This provides us with a unique opportunity to communicate with the eye.

The retinal prosthesis is an effort to provide a sensory substitution, if you will, a replacement for those last photo receptors, another input to the retina, one that can allow us to send visual information through those ganglion cells, down the optic nerve, to the brain, a visual percept that a patient can understand as sight.

The rental prosthesis is a device that has an electrode array. This electrode array is relatively flat and conforms to the inner aspect of the retina, the ganglion cell layer. And that electrical device can stimulate the ganglion cell layer to produce visual percepts called electro phosphines.

The electorate array that we're using has 60 channels. That means that the visual world is interpreted from a camera with a small visual processing unit worn on the belt or via a sling around the chest or shoulder.

That camera image is processed by the visual processing unit, and an electrical signal is sent wirelessly through a pair of glasses to an implanted chip, which is interpreted, and then sent as a series of electrical impulses to the electrode, which then stimulates the ganglion cells in a patterned sort of way.

So this electrical stimulation occurring at the ganglion cell level is spatially and temporally patterned to reproduce a video image. It's a video image with 60 points of stimulation. And these points of stimulation are artificial. They are electrical. They form points of light. And these points of light can be assembled by the patient into something that they actually recognize.

So a patient with retinitis pigmentosa who's lost all visual input, who has perhaps bare light perception, or light perception with projection, or perhaps no light perception, can use the strategies that they've come to learn in the most advanced forms of their vision loss.

SPEAKER 1: And if I move, then I can tell exactly kind of where you're at.

DR. RAYMOND IEZZI: To scan the visual world side to side, up and down, and use these 60 points of visual stimulation to reconstruct a visual scene, the brain is attached to this sensory input. We have a very strong processor attached to this system. And in the form of our human brain, we can interpret what these visual signals are. And patients can often detect where that door is.

SPEAKER 2: Exactly.

SPEAKER 3: That's the right edge of the door.

SPEAKER 1: Yeah, it is vertical.

DR. RAYMOND IEZZI: They can detect where the handle on a refrigerator is. They can identify objects on a table.

SPEAKER 3: So they can get everything that's in there?

SPEAKER 1: Well, I know there's something in there.

DR. RAYMOND IEZZI: They can find their fork and their knife during dinner. They can cook, and determine when their egg is done. They can see their family members.

SPEAKER 2: There.

SPEAKER 1: Yes, there he yes.

DR. RAYMOND IEZZI: They can see how tall they are. They can see where they're moving. They can see their grandkids. Patients with the retinal prosthesis often watch television, even though they cannot make out the objects that are on the screen, they can see motion and they can associate this motion with what they're hearing on the screen. This is valuable to patients. It improves their quality of life.

SPEAKER 3: You just saw your first sunshine.

SPEAKER 1: That was her.

SPEAKER 3: That's true. OK, that's the ray of light coming in through the window.

SPEAKER 1: I know, I know.

DR. RAYMOND IEZZI: They can determine whether the sun is up or the sun is down. It helps their circadian rhythm. And it gives them an additional tool for ambulation. The visual information provided from the rental prosthesis enhances their confidence in mobility. It enhances their ability to use visual information in their day to day world.

SPEAKER 3: You're seeing your reflection in the mirror.

SPEAKER 1: And it's not as bright.

DR. RAYMOND IEZZI: So the retinal prosthesis is a remarkable development. It is capable of providing rudimentary forms of vision in patients who've lost all sight. And while it is providing an artificial vision very different than any form of vision that any of us with sight have perceived, this does have value and has found its way into the daily routine of our patients with the most severe forms of retinitis pigmentosa.

Well, how do we determine who would be a candidate for this? We would need a patient who has had very severe vision loss, to the point of bare light perception. Certainly a patient with hand motions vision would not be a candidate for a prosthesis at this time.

Patients with no light perception are also candidates. But we do have to assess the anatomic integrity of the ganglion cell layer in the inner retina via optical coherence tomography testing and examination. We'd also have to establish that the eye was structurally sound, that there wasn't a severe retinal detachment, and that the sclera itself was capable of handling the surgery. We'd have to establish surgically that the patient could undergo the procedure.

In addition, we need a patient who has the capacity to cognitively interact with the new sensory input from a retinal prosthetic. It needs to fit into their lifestyle. And they have to be motivated to pursue the acquisition of this new sensory input, which for some perhaps would be annoying or difficult to interpret if they did not have certain cognitive skills.

SPEAKER 1: It's like a new toy, a new tool that we need to explore and learn how to use it. And so it's really exciting.

DR. RAYMOND IEZZI: So I would encourage you to consider identifying patients who might have suffered the most severe forms of vision loss due to retinitis pigmentosa. And be mindful of the fact that many of these patients no longer come to the doctor.

So if you have, for example, a family member who's young with retinitis pigmentosa, in taking family history, ask if there are any family members who have retinitis pigmentosa who are older, who might have had more severe vision loss, and reach out to those patients, and let them know that there are potential solutions to improve their quality of life.

SPEAKER 1: It's crude, but it's significant. You know, it's work.

SPEAKER 1: I'm Dr. Raymond Iezzi, here at the Mayo Clinic. Thank you for your time.