DR. RAMIN ESKANDARI: Craniosynostosis, or the premature fusion of one or more of the brain sutures, is estimated to occur and about one in 2,500 births. The fusion of the sutures typically occurs before birth or very soon thereafter. And at times, craniosynostosis can be attributed to certain genetic causes. Those are called syndromic craniosynostosis.

However, in most cases, or nonsyndromic craniosynostosis, you can have one or more sutures involved as well. Our case this time around, was a case of nonsyndromic craniosynostosis. And we used multiple methods of evaluating this patient before we embarked on surgical treatment.

One of the ways that we evaluated this patient was, right off the bat, there was noted asymmetry of the cranial vault. There are multiple types of craniosynostosis. And depending on the type of craniosynostosis, the cranial vault will have different shapes, different stereotypic shapes.

In Figure 1, you can see that there is letter A, B, C, and D. Each of them represent the stereotypic shape of various cranial deformities.

Part A of Figure 1 shows sagittal craniosynostosis with an elongation or scaphocephalic head shape. Part B shows a typical bicoronal craniosynostosis, with a flattened pancake or brachiocephalic head. Figure C shows a metopic craniosynostosis with a typical triangular shaped ridge or pointy head, called trigonocephaly. And figure D shows a nonsynostosis picture for comparison, showing positional plagiocephaly with flattening of one side of the posterior part of the head, mostly from the positioning of the patient in bed.

These four types of typical cranial vault deformities are sometimes hard to differentiate from one another, when they're not this obvious. One of the most important parts of evaluating a skull shape is looking at whether there is mobility along the bony edges.

The next figure is a direct comparison of the suture lines. As you can see, the multiple sutures in part A are all open. And in part B, the midline sagittal suture is closed. The asterisk is pointing towards a ridge that sometimes forms during that fusion of that suture.

Another aspect of the bones of this sort of skull deformity are that when the sutures are fused, they most likely never formed an appropriate suture in the first place. And therefore, the bone across there is smooth. And it looks like there was never a suture to begin with.

Having that bony fusion there prevents the growth of the skull from the underlying brain growth in one direction, i.e. the growth of the skull in a scaphocephalic or sagittal synostosis case, as in Figure 2 part B, shows inhibition of growth in the lateral direction and only growth can happen in the AP or front back direction, hence the elongated football-shaped head.

In our case, we had a child who showed obvious, very severe craniosynostosis with head shape deformities, indicating multiple sutures being fused. In his case, he had not only a sagittal craniosynostosis, but also a coronal craniosynostosis.

When we first saw this patient, he was only a few weeks of age. And at that age he was too young to get imaging to see exactly what sutures were fused. However, we knew that, most likely, he would require surgical intervention. And therefore, we did decide to obtain one set of low dose CT imaging to be able to create a 3D model.

Our 3D model helped us a lot, because we were able to isolate all the different bones and sutures that were abnormal. And in his case, he had a severe unicoronal synostosis on the left and a sagittal synostosis midline.

As we followed him along, he demonstrated that his head shape deformity was progressing really quickly. And we knew that we had to intervene to prevent the abnormal brain growth, as well as prevent ocular problems in the future, with eye socket deformities.

In this patient's case, after seeing the imaging and how thick his bone was and the severity of his multisuture craniosynostosis, it was decided among our group that we would perform a staged operation. The first stage of the operation would release the main sutures that were synostosed and allow his brain growth to develop.

After a certain amount of time, probably about one year, we estimated that we would go back and do the orbital portion and frontal portion of his surgery and reconstruct the cranial vault at that time. However, in light of his young age, we elected to not do all of that in one sitting.

Figure 3 demonstrates positioning of this patient for his second stage surgery. As you can see, the incision line has been marked on the head. It's a wavy incision, to help with the cosmetic regrowth of the hair afterwards.

What we're measuring here is his forehead. It's very important, as you do reconstructions of the anterior portion of the cranial vault, to know how far forward the forehead really should go and the angle of the forehead at the break of the hairline.

Figure 4 demonstrates marking of the incision on the contralateral side, as again a wavy incision is the best incision for cosmesis once the hair grows back. Figure 5 is a head on view from the top showing the incision line and the head being prepped. Figure 6 is going through the original incision.

The way that we make the incisions on these children is to try and prevent as much blood loss as possible. Therefore, we use what's called a Colorado Tip Bovie needle that emits energy directly out of the very tip, which helps with inflammatory reactions around the edge of the tissue, but also helps with obtaining minimal blood loss at the time of incision. As we try and make a hemostatic incision down to the level of the calvarian our goal is to create a subgaleal pocket, over which we can reflect the scalp and minimize bleeding.

As you can see, in Figure 7, we're using a Bovie electrocautery device to lift off the scalp. And in this case, since this is the second surgery, there's a significant amount of scarring on the side on the underneath side of the scalp, because we had already performed an operation. It is, therefore, even more crucial to take your time and lift off the galeal layer, not lifting off the periosteal layer against the skull. The periosteal layer controls a significant amount of bleeding and, if left on top of the skull, can minimize blood loss significantly.

Once the entire calvarium is exposed, you can see in Figure 8 both the pictorial view from a lateral aspect, as well as a rendered drawing that shows the area of interest. The red lines in the drawing indicate the area that we would be removing in order to fix the forehead. The asterisk is a point at which we're going to place our burr hole in order to obtain access.

Figure 9 demonstrates that bony bifrontal area having been removed. And you can see the dura exposed and the piece of bone in our hand. That piece of bone that we removed is going to become crucial in recreating and reconstructing this patient's forehead and anterior cranial vault.

The main reason is because we're able to take that bone, and since the patient is much older, we can split the bone down the middle into two thinner pieces of bone of the same size. This allows us to literally double our bone availability to reconstruct the patient's forehead. This is being shown in Figure 10 using a reciprocating saw.

Figure 11 demonstrates, so to speak, a jigsaw puzzle being put back together. We're using various pieces of bone that we have created from the patient's original craniectomy that we took off at the surgery time. And we're organizing the pieces into various areas of the skull. We're using a retractor underneath the bone, but over top of the dura to protect the dura from the drill, where we're going to fasten the bone edges together.

Figure 12 is a representation of the patient's skull before the first operation and after the first operation, a year later. As you can see, this is a top view of the skull showing the synostosed suture in the left-hand picture with the coronal and sagittal synostosis, as well as on the right-hand picture after the release of those bones.

What you can see is that the head has now been able to grow in a much more rounded manner. However, there is still a significant frontal bossing on the right side, as a compensatory mechanism of the area where the brain can grow versus the left side where the brain could not grow.

Another thing you'll notice is that there are still some cranial defects from the area that was released. This is OK, given the fact that we knew we would be going back and completely reconstructing that skull in the first place.

Figure 12 shows same two skull models but from an anterior view. You can see the significant ocular deformity caused by the pulling of the coronal synostosed suture, because the brain is trying to grow and it can only grow onto the right side.

The left side it looks like it's being pulled backward. But in reality, it just hasn't moved and the right side is being pushed forward. You can see how the ocular deformity is significantly corrected a year later, just from removing the craniosynostosed sutures of the left coronal and the sagittal suture.

Figure 13 demonstrates a more complex portion of the case where we're reconstructing the anterior cranial vault. And because we have to correct for the lack of growth on the left side, we have to advance the cranial vault forward. This orbital and frontal advancement is performed by putting strut grafts of the patient's own bone in between the new bony pieces and securing them in place so over time they can fuse and complete the skull.

You can see, based on the drawing in the bottom right corner that there will be a gap between the frontal bone that we've reconstructed and the patient's own bone in the back there'll be two strut graft that will hold it in place. But what's important to note is that the dura underneath also has osteoblastic properties and the bone can grow, not only from the edges of the existing bone, but also from the dura on up and completely fill in any gaps.

It's important to note that, in general, cranial facial surgery, including skull reconstruction, has been associated with large volumes of blood loss and, many times, need for multiple blood transfusions. We've attempted to try and decrease this potential side effect. It has complications, including severe side effects and complications of lung injury.

By rendering our CT scan's, a useful preoperative and intraoperative tool, we've created 3D printed models of the patients' skulls before we do any kind of open surgery. This allows us to not only review the surgery we need in 3D, but to have a hand held exact replica of that patient's skull. We can even cut it, make drawings on it, perform the surgery, so to speak, before we actually get to the patient.

This has significantly improved our surgical times and decreased our amount of blood loss, because we're not wasting time in the operating room manipulating the bones or trying to come up with a plan. This has all been premade. And it's been vetted through the 3D printed model. Planning surgery using 3D models of the patient's skull and eventually practicing on those models can significantly reduce operative times, blood loss, and need for transfusions.