I'm happy to have the opportunity to share with you our experience over the last 10 to 15 years with pediatric skull based surgery and the application of the endoscopic approaches for this. So as we are all well aware, the use of the endoscopic approaches has really rapidly expanded over the last 15 years and the technological advances have allowed us really better visualization and has also given us the opportunity to decrease the size and the instrumentation. If we look at our pediatric experience between the years of 1999 and 2005 we had done or treated a total of 25 patients and we looked at the complications in those patients and presented that in an article that was published in 2007. Between 2006 and 20011 we treated and an additional 118 patients and at this point we have now done or by the end of 2011 had done over 170 procedures in the pediatric population. And in this case we are looking at patients under the age of 18, or 18 and under.

Much as you have already heard and seen throughout the course so far the endoscopic approach gives us access not just to the sellar region and areas nearby the sellar region but also really gives us access to the anterior skull base, whether that's through a trans plenum approach or a trans cribriform approach we also have access to the frontal sinuses as well as access to the posterior fossa coming through the clivus and all then all the way down to the craniocervical junction. We also much as we have been able to do in the adult population have been able to extend this out of the midline and into the coronal plane, giving us access into the pterygoid regions, the infratemporal region and really giving us a wide variety of tumor types and locations which we can access through a minimally invasive approach.

If we look at the split of the approaches that we have been using you can see that about 25% of our approaches involve the anterior skull base; these have typically been dermal sinus tract and encephaloceles as well as some dermoid and epidermoid tumors. About a third of our cases are really trans sellar and I would the majority of those represent craniopharyngiomas as well as other pituitary lesions. Almost a quarter of our approaches have included transclival approaches, this has included some intracranial posterior fossa pathology as well as a number of chordomas. About a third of our procedures now extend into the coronal plane, this includes the J&As and as well as we've done some rhabdomyosarcomas and a number of other lesions and really only about 10% of our cases have been limited to the sinus and orbit.

If we look at the pathology that we are primarily seeing, and these would be sort of our most common pathologies, about 20% of our procedures still include nasal angiofibromas. About the rest of them are sort of split in the 10% range with about 15% craniopharyngiomas, about 10% of chordomas and Rathke's cleft cyst and again pituitary tumors.

So the things that I think have been really key in applying this in the pediatric population has included septal flaps giving us the ability to do a vascularized flap. Also we have utilized – I don't know if that's going to work. We'll wait until it falls over then we'll all laugh. Three we go. So the other thing that we have utilized is in some cases we have, we have included a sublabial approach. The sublabial approach has really been restricted to those cases where the nares were so small that

we didn't feel like we could place adequate instrumentation. But again this has been really reserved for the very smallest of children who have been involved.

The other thing is in the pediatric population hemostasis must be meticulous. When we are operating on the 5 year old we can probably tolerate a 500 cc blood loss, whereas if we are operating on a adult we may be able to easily tolerate a 1 liter more blood loss. For this reason there has been some instances where we have chosen to do staged procedures where if the bony access has had a sufficient enough blood loss that we felt that we should go ahead and stop and come back another day. And this might seem a little bit strange because for the most part when you are working in adults you have well pneumatized sinuses and you don't have to do significant drilling simply to really the sella or the clival region, whereas in the pediatric population especially when we are working in children under the age of 10 who have very poorly pneumatized sinuses we may actually drill through a centimeter or more of bone before we even reach the sella. And so unless you are extremely meticulous about controlling the blood loss during that you then really can run into a problem of a significant blood loss before you've even reached the pathology for which you have set out to treat.

The other thing that we have felt very strongly about is the team approach. We looked at a number of models when we were first deciding to actively extend this into the pediatric population and from the standpoint of sort of quality care for children we felt that having the adult team do the surgery and drop them off at the pediatric hospital was less optimal than having the pediatric neurosurgeon

involved in the procedure. But we also felt given the simple numbers and the volume of procedures and the learning curve associated with these procedures that having a pediatric neurosurgeon be the only one involved with the ENT colleagues was also probably suboptimal and that by creating a 3 person team that included an adult skull base surgeon, a pediatric surgeon and the ENT component to this really gave us the optimal approach to this.

I think also patient selection becomes very important and I think one of the things that we certainly see is that there are times when we see lesions that will appear to be benign lesions and rather than going in for an aggressive surgery to actually confirm a diagnosis we have chosen a wait and see approach. Certainly we've had a number of nasal pits with dermal sinus tracts that we have identified in children who are newborn who we have chosen to wait until the children are 2 to 3 years of age before treating. So we have taken an approach sometimes where we have watched the lesion rather than rushing to surgery.

So I want to take the opportunity to give you some perspective on the variety of lesions that we have had the opportunity to treat. We are going to first look at some of the sellar lesions. This was a teenager that presented with what was felt to be a Rathke's cleft cyst with intractable headaches. So this is the video from that, that particular surgery. The dura has already been opened, the tissue is being opened at this point. This was a very mucinous lesion, this patient has resolution of their headaches, postop imaging showed that the cyst had been decompressed and on follow-up we did not have evidence of recurrence of cyst. So as would be typical for Rathke's cleft cyst we did not do

a closure at the end of these procedures but simply leave them open so that they can drain into the sinus. And again this is just making sure that the material has been removed. And I have to stop clicking so fast because otherwise I'm just going to go past everything that's on the slides. So here is the postop imaging, you can see that the cyst has been decompressed.

When we look at sellar lesions we've also treated a number of sellar craniopharyngiomas. This was a young lady who was from the northeast. She had this preop imaging. She visited 3 other institutions, all of which recommended an open craniotomy for treatment of this lesion. This mother had read about the endoscopic endonasal approaches and actually came here for an evaluation to see if her child would be a candidate. This is her postop imaging now – she is now 4 ½ years out from her resection. I think these images were done right about the 4 year mark. You can see that in this child there was residual pituitary tissue that was left behind. She appears to have a stock in place. She has actually been on hormone replacement, although she is not on complete hormone replacement. She is currently off of DD ABP and she has continued to do well and we are fortunate enough to have her long term follow-up films because they drive from New Jersey once a year to come down to see us.

We also have treated some things that were not quite so obviously cystic lesions. This was an exceedingly calcified lesion, in fact there was debates as to whether or not it was a craniopharyngioma, and you could see the heavily calcified lesion here. This tumor was actually quite difficult to remove because it was sort of notched around the third nerves and so every time we

would wiggle it we would be wiggling the optic nerves or the third nerves, and we literally resorted to sort of chiseling off pieces bit by bit so holding it still with one piece and biting pieces off. We were quite happy with her postop imaging. This was sort of mediate postop imaging. We did not use a balloon in her. I believe she was probably about 5 at the time and we ended up packing her. Her packing was removed in the OR because sometimes young children can be a challenge to remove their packing in the office. This was her one year postop imaging, you can see her vascularized flap. You can see the relatively empty sella. Over the next year she developed an area where she had some enhancement, this has now been stable over 3 years. She is now 4 years out from her lesion. She had a third nerve palsy immediately postop that recovered over 3 months.

Now for other lesions that extend anteriorly we can include the trans planum approach to handle larger lesions. This is another child who presented with a craniopharyngioma. You can see they had these two large cysts extending up into the third ventricle. This was the calcified or the solid component, he had a CSF cleft that was here. It was unclear to us whether or not he had residual pituitary tissue prior to the surgery. To reach this and to make sure we had good visualization of the optic chiasm we did both a trans sellar as well as a trans planum approach, and again you can see that his sinuses are not well pneumatized, so you can imagine we did a fair amount of drilling before we actually opened. This is after we had opened the dura, you can see there is some residual pituitary that's compressed posteriorly here. This was after we had released the CSF from here. You can see up here at the top is the optic chiasm and here you can see the tumor. We have a vessel that we felt was most likely also feeding the chiasm that we were eager to preserve. Here we are

beginning to open the arachnoid over this, now we've actually opened all of that arachnoid preserving that vessel that we felt was most likely feeding the chiasm, beginning to open into the tumor. You can see here we are removing solid tumor. In this particular case once we had removed that solid tumor in the bottom that opened up into the two upper cysts allowing them to collapse, continuing to sort of take that down.

And what I'd like to show you is, and this is a philosophy that we have adopted, we feel very strongly that stripping the membranes off of the cysts from craniopharyngiomas that extend up into the third ventricle dramatically increases the likelihood of hypothalamic injury and so we will not infrequently leave a cyst wall that is adherent to the walls of the third ventricle behind so you can see this rim of enhancement. This was the pituitary tissue that we had left behind. At one year he had a recurrence, not from what's left in the third ventricle but down here where the pituitary tissue was. He had a second surgery at which time we removed all of the residual stock and pituitary tissue.

So there on occasion you will see larger lesions or lesions extending into the posterior fossa and we can add a transclival approach to that. This was a young child, at the time they came to us I think it was just before his 4th birthday. He had already had 2 open craniotomies in his home country, and they had not been successful in resecting his craniopharyngioma. He now had this large cyst that extended into the posterior fossa. The family visited multiple places throughout the world trying to find a place to come. You can see also he had this cyst that extended up into the third ventricle. The two prior craniotomies had left a fair amount of scar along the carotid from the site of the approach

but we were able to resect the cyst going up into the third ventricle. You can see again we did leave a little bit of enhancing tissue that was up against the wall. It was very densely adherent to the wall of the third ventricle there. And we can see that we were able to decompress that posterior fossa cyst and this is just the coronal view. This child went on to return to his home country. It was anticipated that he would likely have some recurrence or repeat appearance of a cyst given that he had multiple surgeries and we knew there was an area that we had left behind, so had setup and made arrangements for the child to have gamma knife should there be evidence of any recurrence. His 3 months scans were clean, his 6 month scans he had a small recurrence of a cyst. He underwent gamma knife. He is now a year and a half out from the gamma knife and these are his most recent follow-up films. You can see that he has no large cysts in the third ventricle, he does have a little bit of enhancement out here along the posterior fossa but has not had any recurrence of his cyst.

The only complicated thing about this particular patient is that when he first went back to India he had a shunt malfunction which was not addressed immediately because they actually decided to send all the films to me first to have me review the films to determine whether or not he was having shunt malfunction, which probably delayed his shunt revision by about 2 weeks. At this point his – they have been comfortable managing him without us reviewing the films.

I'd like to give you a couple of examples of things that we can do at the craniocervical junction. This was a young lady who had congenital scoliosis prior to her thoracolumbar fusion. The orthopedic surgeon taking care of her imaged her entire spine. You can see some of the segmentation

anomalies and fusions she has as you enter the upper thoracic spine. She had this basilar invagination which we treated with an anterior approach taking down the dens all the way to the body of C2 and I'll show you on the MRI in just a minute, we also did a partial Chiari decompression of this sort of long spit that was sticking down giving her a nice decompression at the craniocervical junction. Just to give you an idea of what her MRI looked like before and after you can see she has a hollow cord syrinx filling the canal. She really has significant compression of the brain stem here. Postoperatively you now have a CSF space that's visible anterior to the brain stem up here, you can see that her syrinx has collapsed. She went on to have her thoracolumbar fusion, she is doing well. The only down side that she reports from having had the endonasal approach for the decompression anteriorly is that she was a trumpet player beforehand and from a little bit of stretch on her soft palate she had difficulty maintaining air pressure afterwards and then was actually found to have a submucosal cleft that had never been identified. So she switched from the trumpet to flute and continued to play in the marching band.

This was an unfortunate young lady who came to us. She had been treated for a Chiari malformation elsewhere, and it was felt that she had some cervical instability. Unfortunately at the time they did her occipital cervical fusion they used BMP to aid in the infusion and she ended up with a fusion mass that was much worse than her original Chiari. And you can see this large bony fusion mass and you can see that her cord is quite compressed. She's also – she is a Klippel-Feil, you can see some of the abnormalities are visible although not super clearly here. Because she was so – had so recently had her posterior cervical fusion we debated what was the best approach to this and after discussion

amongst the pediatric neurosurgeons it was felt that perhaps giving her an anterior decompression would be the best for her rather than disrupting her recent posterior fusion. So we approached this from an anterior approach, you can see now we've given her a nice CFS space, she's actually not tight here, there is a little bit of an artifact but she actually has a decent CSF space and if we look at the bony decompression again you can see we've opened up this nice space here and as so she went on to do well. She did because she was a Klippel-Feil and was essentially fused up to the level of her one mobile segment ended up developing hypermobility at that segment. We went ahead and extended her cervical fusion and she has actually done quite well now and has been pain free for over a year.

So you know again we really have been able to approach a wide variety of lesions in the midline, including the anterior approach which we haven't really looked at. So this was a young child at the time she came to us. She – I think it was just after her second birthday, you can see she has this encephalocele that is here. Family reported that really other than some nasal stuffiness that she was not symptomatic. Looking into this as we open the nose, first looking in the nose you can see that there is this weepy thing here, a large mass. Now in retrospect when I talked to the family when she came in for her first postop visit the family said this is great, we were able to throw away all the Kleenex boxes that were in every room. So in fact she had been leaking her entire life. They just hadn't recognized that it was rhinorrhea. Here we have now opened that sac and we are now shrinking back the neural tissue, continuing to shrink that back. You are beginning to see the bony edge all the way around. Now you can with sort of a pulled back view you can see we've exposed

bone circumferentially around the defect and shrunk all the tissue back. We put a small free mucosal graft over this. This is her postop image. There is no more encephalocele, we see some changes from the surgical approach and she has continued to do very well and is almost 2 years out at this point.

This next case again looking into pathology from the anterior fossa at the time of this imaging this was a 10 year old. He was involved in a bicycle accident, he did over the handlebars after hitting a wall. Unfortunately on his CT scan a large mass was identified and so we had this large intradural dermoid. This is just some video of resecting on the intradural dermoid. In this case we are now opening the dura, you can see the pearly white material and using suction being able to now take this off, you begin to see neural tissue back here. And one of the keys about this is as you are resecting this one of the things that can begin to happen is that it all will begin to collapse down, so great care really needs to be taken in trying to make sure that you don't allow the cavity to collapse, leaving a segment or an area of tumor that you've been unable to resect. Again here is just an example of slowly removing the tumor, beginning to expose the optic chiasm. And we are going to go ahead and keep going.

So this is his postoperative imaging, you can see we've left him with a large defect here with a vascularized flap. You'll see on the postop imaging there is this small area of T2 signal that was left behind. It was unclear whether or not this was just CSF or tumor on the DWI imaging. The area was small enough that it was unclear what it was. He did exceedingly well but at 6 months postop

presented with acute meningitis, was presumed that he had a leak, he was taken to the OR and imaged, or actually visually inspected. There was no evidence of a leak and in fact what he had developed was an abscess in this general area that was stereotactically drained. He's now 2 years out from the treatment for his abscess. Actually he's almost 3 years out at this point and continued to do well beyond that point.

Some examples of things that we have been able to do extending out into the coronal plane, this is a young child who was about age 5 and he presented with severe V2 pain; imaging identified this rhabdomyosarcoma which was biopsy proven to be a rhabdomyosarcoma. The typically treatment would be chemo and radiation but due to the severe pain that he had we were asked if we would consider a resection to see if we could provide him with palliation of his severe pain. You can see that there is a small knuckle that extends into Meckel's cave. These are his postop images. We felt that we had an excellent resection, we have removed that area that extended into Meckel's cave and that just sort of popped out leaving the arachnoid intact and was covered with a vascularized flap. From a palliation standpoint there was excellent (inaudible). He had complete resolution of his pain following surgery. He did go on to have a course that would be expected from a rhabdomyosarcoma with intracranial extension, but went on to survive for another 2 to 3 years.

The other question that I just wanted to show you an example of this, I talked about the fact that using the vascularized septal flaps has been very important for us. On occasion we have had some dural defects that have really occurred at the level of the frame and magnum, we had a patient who

was a multiply recurrent chordoma, had previously undergone proton beam therapy and he had a CFS leak that was really all the way down at the frame and magnum. We did not have great coverage of that with a nasal septal flap and in order to treat a recurrent leak from that we did a pericranial flap so it was brought in through a slot at the level of the frontal sinuses and you can see this is up close having laid it all the way down to the frame, below the frame and magnum. This is sort of the pulled back approach, you can see the bottom of the flap. Our gel foam here this now covering the sella area, this coming anteriorly really allowing us to bring a vascularized flap all the way down to the level of the frame and magnum. This worked very well and he did not have a persistent problem. We actually subsequently have done another surgery on him, so we had an opportunity to see how well this had healed, and actually had provided him with a very nice vascularized coverage of his bony defects.

So to review the complications we've seen in the pediatric population, we've had no deaths. Out of those 133 patients we have had 3 patients who have developed permanent cranial nerve palsies. We've had probably 8, about 8 transient nerve palsies that have recovered. There have been a total of 14 CFS leaks since the institution of using the vascularized flaps or the septal flap, our CSF leak rate has dropped significantly. We seen a total of 6 cases of meningitis and as I mentioned we've had some abscesses that have presented in a delayed fashion. This has been, certainly has provided us with an effective approach for a variety of pathologies in the pediatric population with a low morbidity. Certainly we need to continue to follow these patients. I think most of our patients now still have less than 5 years of outcome, so determining whether or not acquired meningoceles or

encephaloceles becomes a problems this has not been a problem that we have seen at this point. Also looking at quality of life, one of the things that we have not seen is we have not seen any significant growth abnormalities in the young children we've treated but again our follow-up is still limited in time. And the other thing is looking at disease control in the – over the long term and comparing that to traditional outcomes, or approaches.

I'd like to take a moment to thank Dr. Snyderman, Dr. Gardner, Dr. Wang and Dr. Fernandez-Miranda who have been my partners in this and Maria our current Fellow as well as Srinivas who is one of our medical students who helped put together the most recent retrospective review of all the patients. And I'd be happy to take any questions at this point.