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STRABISMUS CASE

1. Could we eventually see the outcome of the first case? the last video 2 months after Botox?

KRD: Yes, I hope MCGN can provide more information on this

SK: At two months, the abduction had improved in the left eye

2. Was there a neurological clinical exam from the paediatricians? This could be helpful too.

KRD: Not that I am aware of. But good point to highlight the importance of a multidisciplinary team approach.

AMM: I agree that a multidisciplinary team approach is needed to manage a case like this. The management of the case would vary depending on coexisting additional neurological findings.

MCGN: Yes, the child was also seen by the paediatricians.

SK: I believe we should refer a case of paediatric sixth nerve to a neurologist for evaluating other cranial nerves as well. We as paediatric ophthalmologists can examine the fourth nerve, sensation from fifth and seventh by other facial features.

RB: I always prefer to get a complete neurological evaluation performed in such a case, it is useful both for diagnostic and therapeutic stand point in case there are concomitant neurological associations.

3. Would they not have a head tilt and be much more ill with Atlanto-occipital lesion?

KRD: Head tilt in atlanto-occipital lesion will be probably one of the presenting features in in this type of injury, however, depending on the degree of injury occurred to the child, there will be a wide variable of clinical signs, one of them sixth nerve palsy due to a stretch injury to the nerve. Please see Neurosurg Focus 14 (2): Clinical Pearl, 2003 for various presenting signs of atlanto-occipital injury in children.

AMM: Similar to the KRD's response, there can be highly variable clinical signs. Even if there was a coexisting ipsilateral CN 4 paresis, it can be masked by the CN 6 paresis. Sometimes the CN 4 paresis will improve, as was seen in this case, only to reveal the non-resolving CN 6 paresis.

SK: Atlantooccipital dislocation is probably more common than realized, especially in the paediatric population. Fourth nerve palsy can be picked up by excyclotorsion as well on fundus

RB: As mentioned above Head tilt is a very variable sign and can be present in such a case. Invariably it is hard to gauge at 4 month of age and excyclotorsion of the fundus is a more definite sign of concomitant 4th nerve palsy.

AF: Head tilt is a sign that could be present in different pathologies. Fundus excyclotorsion is more related to 4th nerve palsy.

4. Is it possible to have Atlanto-occipital lesion without neurological sequelae? Do they all need plate?

KRD: I don't know. Perhaps MCGN who was involved in this case can elaborate on this.

AMM: I think the decision for neurological imaging depends on how long after injury the child presented for ophthalmological evaluation and resources available where the child's care is being provided. In most settings, an acute onset CN 6 paresis in a child of this age after trauma would warrant neuro-imaging.

MCGN: I would recommend this reading: Astur N, Sawyer JR, Klimo P Jr, Kelly DM, Muhlbauer M, Warner WC Jr. Traumatic atlanto-occipital dislocation in children. J Am Acad Orthop Surg. 2014 May;22(5):274-82. doi: 10.5435/JAAOS-22-05-274. PMID: 24788443.

SK: Any infantile sixth nerve palsy should be investigated with neuroimaging to exclude slow growing tumours. From an ophthalmic point of view, they might resolve but the aetiology has to be established.

RB: I believe there can be variable presentations with atlanto-occipital lesions, it depends on the severity, exact location and chronicity of the lesion.

NON-STRABISMUS CASE

5. Since there was a Descemet dehiscence seen on UBM. Could you consider doing a DMEK prior of the PK?

KRD: Good point. We could probably consider doing a DMEK prior of PK, however there has been no sufficient scientific evidence to support the superiority of DMEK in paediatric population. There was a case series published in Cornea: September 16, 2020 - about the superiority of DMEK in treating endothelial dysfunction in children, however this procedure was considered more difficult.

PS: No, in this eye an endothelial transplant would not be helpful. The cornea is not abnormal from purely endothelial dysfunction or glaucoma – it is structurally abnormal with abnormal stroma and scarring. This is evident on the UBM showing the posterior corneal pseudocyst. "Trying" an endothelial transplant in these eyes is not only difficult but could be dangerous. (a) There is no view of the anterior chamber. (b) There is a serious risk of lens touch and cataract. (c) It would delay clearing the visual axis, which is time critical in these infants who have dense amblyopia already.

SK: Very good option. But outcome of DMEK in paediatric population is understudied.

FC: You can try but is difficult to do it, particularly in this case, just because you can't see good enough. It will be difficult to unfold the graft and you can touch the lens. The patient has developmental abnormality affecting the corneal stroma as we can see in the UBM so in my opinion a Penetrating Keratoplasty was the best option.

6. Role of Digital IOP?

KRD: In cases of corneal opacification, we simply cannot always rely on tonometers as abnormal central corneal thickness can affect IOP reading. For further reading on the comparison of IOP measurement tools in paediatric population, please see Survey of Ophthalmology 2019;64(6):810-825.

PS: In structurally abnormal corneas, "digital" tonometry may be the most accurate way of measuring IOP. All devices rely on some assumptions about corneal biomechanics.

SK: Very useful in scarred corneas

RB: Digital tonometry is a very useful method of assessing IOP in patients with structural corneal abnormalities and sometimes can be the only method to estimate the IOP in such cases.

AF: Digital tonometry is useful in “trained hands”

FC: Very useful in cases of corneal opacification. We have to train “our hands” in normal and abnormal subjects.

7. Should we get used to digital IOP in theatre in 'normal' subjects?

KRD: it would be helpful to have a handful of experience in measuring digital IOP in normal subject to be able to compare to those with high pressure.

AMM: Practicing digital IOP in theatre is helpful for a starting point in honing this skill. It is also helpful to digitally assess IOP in the clinical setting with an awake child to gain familiarity with doing this in a less controlled setting.

PS: Definitely.

SK: Knowing and practicing digital IOP taking can be very helpful in clinics as well as operating rooms in scarred corneas.

RB: Absolutely, I always assess IOP digitally after checking it with a ICARE in the clinic setting as in my “fingers” it is very reliable.

AF: Sure

FC: Absolutely.

8. Under what type of anaesthesia was this IOP measured? Was it at induction?

KRD: General anaesthesia, at induction.

PS: General anaesthesia, at induction.

SK: At the time of induction in general anaesthesia.

FC: General anaesthesia, at induction.

9. Could an IOP of 20 mm Hg make the cornea opaque?

KRD: Not that I know of.

PS: No, the pressure is not actually 20. And the corneal opacity is not just from pressure but also developmental abnormality affecting the corneal stroma.

SK: Not to my experience.

RB: It should not if the Endothelial layer of the cornea is intact.

AF: No

FC: No

10. Did you try to image anterior segment with OCT/UBM?

KRD: We did

PS: Yes, we did. These are shown on the subsequent slides.

SK: Yes, UBM would be more informative to look at the lens and ciliary body.

FC: Yes, UBM and Anterior OCT were very useful to illustrate this case.

11. What is the IOP variation under general anaesthesia?

KRD: Typically to avoid false reading in IOP under GA, measurement should be done at induction.

PS: It depends on which agents are used. Succinylcholine and ketamine increase IOP, hypercapnia increases IOP. Laryngoscopy, relative airway obstruction and inadvertent pressure on lower eyelid/globe with the anaesthesia mask can increase IOP. Most other anaesthesia agents reduce IOP. Inhalational anaesthesia rapidly reduces IOP.

SK: IOP will change after intubation. So best is to measure just before/at the time of induction.

FC: It changes after intubation.

12. Diode in an eye with abnormal anatomy: how would it be directed at the CB? Destructive and pro-inflammatory? Or is there place for TSC in management?

KRD: Typically, TSC in paediatric glaucoma can be used to temporized pressure control before proceeding to other modality of management such as GDD. The location of CB may be able to be visualized under UBM guidance or using a linear ultrasound. UBM has been very helpful during my fellowship in a series of paediatric ophthalmic disorders, some of them with congenital corneal opacities and high intraocular pressure due to anterior segment dysgenesis. TSC guided with UBM was performed in these patients prior to other managements.

PS: Cyclodiode has an important place in management of paediatric glaucoma. It is directed at the ciliary body, and yes, it is destructive and pro-inflammatory. Often, it needs to be repeated. In abnormal eyes where clinical examination and transillumination do not guide location of ciliary body, I use this technique using UBM water bath: Way AL, Nischal KK. High-frequency ultrasound-guided transscleral diode laser cyclophotocoagulation. *Br J Ophthalmol.* 2014 Jul;98(7):992-4. doi: 10.1136/bjophthalmol-2014-305163. PMID: 24692746.

13. Did you choose to transplant the "better" eye (in terms of glaucoma)?

KRD: Perhaps PS can elaborate more on this, but I think it is common sense to try to save the better eye first.

PS: These are very abnormal eyes. Expulsive haemorrhage is a real risk. I tend to recommend transplantation of the eye with less potential first, and see if there are any major intraoperative complications. There is less to lose if there is a complication. If all goes well, the better eye can be done soon after.

SK: Yes

FC: It depends. If both eyes have good visual potential I go with the less potential first. But if I know that I have one chance I will go to the better eye. Finally, I share with both parents the decision.

14. With what instrument do you follow intraocular pressure in patients with new corneal transplantation?

KRD: In my opinion, iCare tonometer be an easier tool for measurement especially in the clinic

PS: In clinic – iCare and digital.

SK: icare can be very useful

RB: ICare, digitally and rarely with tonopen.

AF: ICare and digitally

FC: ICare, digitally and with tonopen.

15. Parth, what donor cornea did you use? What is the ideal age of donor?

PS: We usually get the youngest donor available on the day from the Eye Bank. Ideal characteristics of donor tissue –

- Donor age < 30 years with cell count > 2700
- Donor age 30-50 years with cell count > 2900
- Age > 50 only if cell count > 3000
- No dropout, no polymegathism, no scar unless in periphery with central clear zone > 8mm diameter.

FC: As younger as you can, good cell count.

16. Does developmental progress improve in infants even if graft fails in children?

KRD: Paediatric keratoplasty is not easy to manage postoperatively, there is a high risk of graft failure, but it is still the mainstay treatment of choice in congenital corneal opacification.

PS: Definitely. The child has some functional vision now that the visual axis is clear. This will continue to develop over time, and aid in the child's general development. Graft failure can be managed medically and surgically. Without timely initial corneal transplantation and clearance of the visual axis, the prognosis would be very poor.

SK: Even if the pre and postoperative course is cumbersome in transplants in the setting of glaucoma; Every child deserves a chance of corneal transplant.

FC: Yes, sure. We have to give them a chance. Corneal Transplant can help in the development of the child.