ELECTIVE (SSC5a) REPORT (1200 words)

A report that addresses the above four objectives should be written below. Your Elective supervisor will assess this.

I decided to do an elective in craniofacial surgery as I wanted to further understand the management of abnormalities which arise from aberrations in the developmental processes of the skull and its associated soft tissue. Commonly encountered conditions include cleft lip or palate, cranial vault abnormalities including craniosynostosis and plagiocephaly, and vascular malformations - conditions which are not frequently encountered on routine medical placements. Furthermore, I wanted to gain an insight into the workings of members of the multidisciplinary team involved in the management of these complex conditions at a highly specialised unit.

I was fortunate to have had my elective at Great Ormond Street Hospital (GOSH) which is one of the four highly specialised centres in England to provide services to children with craniofacial abnormalities.[1] GOSH is one of the largest craniofacial centres in the UK and treats 250 patients a year with craniofacial abnormalities and performs approximately 200 operations annually.[2] Although the most commonly encountered facial deformity in the UK is cleft lip and palate which affects around 1 in 700 babies nationally,[3] the majority of my time at GOSH was focused on patients receiving treatment for craniosynostosis. This was most likely influenced by me undertaking my shadowing with the neurosurgery department at GOSH whom form part of the craniofacial team but are predominantly involved in the management of calvarial vault abnormalities. Craniosynostosis is a rare defect and occurs when there is a premature fusion of the bones forming the cranial vault. This can be isolated or multisutural. Sagittal synostosis is the most common form of craniosynostosis affecting 55-60% of cases.[4] The incidence of craniosynostoses is reported to affect approximately one in every 2000-2500 births,[4] however there is variation in the literature of exact figures globally.[5, 6] A common theme is the recognition of the rising incidence of craniosynostosis with time,[5-8] this may be due to increased awareness amongst health care professionals and reporting amongst parents.[7] However, maternal factors (e.g. maternal birth history, smoking, race and ethnicity) and environmental factors (e.g. in utero teratogen exposure, foetal positioning and folic acid supplementation[8]) could also contribute to this rise.[4]

Operative management for craniosynostosis is broadly classified into calvarial vault remodelling (CVR) and minimally invasive methods of suturectomy with spring assisted cranioplasty (SAC) or helmet therapy.[4, 9] SAC was a procedure which has been adapted by surgeons at GOSH since April 2010 specifically for correction of sagittal craniosynostosis.[10] The procedure works on the principal of distraction osteogenesis where springs are placed in a bone gap formed following osteotomy of the prematurely fused suture.[10-12] Gradual expansion allows for correction of head shape. CVR procedures are visibly more complex with regards to operative skills and pre-operative planning required. CVR involves reorganisation of cranial bones in a jigsaw like manner to correct for bony prominences produced by craniosynostosis. I was amazed by this operation and admired how plastic surgeons and neurosurgeons integrated their skills to architecturally refashion the calvarium. Furthermore, the variety of materials and instruments, especially the use biomaterials like bone 'salami' (a composite morsellised bone mixed with fibrin glue)[13] to fill defects in the cranial vault and aid remodelling of bone was eye-opening. Although I did find the CVR procedure more intriguing, I learned not to get distracted from the benefits of minimally invasive SAC. Here I was amazed by the speed of the operation and the fact there was visibly less intraoperative blood loss compared to the CVR procedure which is analogous to what has been reported in the literature.[10, 14-16] The advantages of SAC was echoed following my conversations with surgeons who applaud the minimally invasive approach with regards to efficiency and reduced perioperative burden for children who can often be discharged the following day. Overall, a mix of age at presentation and form of craniosynostosis are the major indicators for type of intervention chosen by surgeons.[9]

Prior to this elective, I conducted an audit with the craniofacial department at GOSH to monitor the long-term outcomes of infants who underwent operative intervention for sagittal craniosynostosis. During the process I learned about the diagnostic workup and follow up of these patients following intervention. Diagnostic work up would often consists of referral from local centres to GOSH. A surgeon in the craniofacial team would then perform a systematic physical examination and if necessary, order radiological investigations to aid diagnosis and pre-operative planning. Following initial assessment, operative intervention would be offered if appropriate. Often the indication for operative intervention in sagittal craniosynostosis would be head shape concerns reported by family members with a small minority having clinical concerns (e.g. raised intracranial pressure (ICP) or visual concerns).

Following intervention children would be offered routine follow up by craniofacial and ophthalmological teams up to a minimum age of 7 where any concerns regarding raised ICP, neurodevelopmental or aesthetic outcomes could be detected and acted upon. If necessary, referral to psychology or speech and language teams was also done. This was a holistic approach to care and it required integration of teams and sustained input of resources and finances to maintain. There are obvious limitations which can be present in developing countries which may lack well supported health care systems with limited funding, skilled personnel and resources to efficiently carry out such services.[17] However, the situation is evolving; centres across the world are documenting protocols adapted to recourses and operative experience at their institutions.[18, 19] Larger emphasis is placed on pre-operative planning in resource limited countries to mitigate complications, but it is still early days with regards to long term follow up protocols which can be found online.[19] Therefore, I believe greater transparency and collaborative learning between institutions may aid standardised, safe and efficient care to be given to those in less fortunate positions than us.

The primary aim of intervention in most cases of craniosynostosis is to correct abnormal head shape, additional benefit may arise in expansion of the cranial vault improving cerebral perfusion and reducing intracranial pressure in certain cases. GOSH has helpfully published craniofacial clinical outcomes in the form of 'Patient Reported Outcome Measure's' on their website which can be used by parents.[2] I thought this was important because it has been found one of the biggest burdens faced by parent's pre-operatively is understanding what to expect following intervention.[20] Interestingly, GOSH found that the biggest motivators for surgery to be head shape, forehead shape and reducing the chance of bullying. Six weeks following operative intervention, there was improvement reported by patient's families in these domains with over 90% of parents/carers reporting surgery to be worthwhile. The burden craniosynostosis overall depends on associated comorbidity and whether there is an associated syndromic cause. Most cases I witnessed were of non-syndromic craniosynostosis. One thing I could have done more to explore the burden of craniosynostosis would have been to attend more follow up appointments offered at GOSH. This would have given me the opportunity to communicate with families directly and learn about any continuing concerns if any they had after operative intervention. Overall, the neurocognitive, behavioural and psychological implications of a diagnosis of craniosynostosis is an extensive topic of its own which varies according to type of craniosynostosis.

I thoroughly enjoyed my time at GOSH with the craniofacial team. Experiencing the work of world specialists at a state-of-the-art facility was a great learning opportunity. The placement consolidated theoretical knowledge I had gained and enabled me to witness the real-life workings of the department and the operative skills of world leading surgeons.

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