PLASTIC SURGERY

Elective Report, SSC 5c
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Objectives set by Barts and The London, School of Medicine and Dentistry were:

1) Describe the pattern of disease/illness of interest in the population with which I worked and to discuss this in the context of global health
2) Describe the pattern of health provision in relation to the country in which I worked and to contrast this with other countries.

My own objectives were:

1) To learn about the incidence and specific features of some syndromes that involves craniofacial deformities and to get insight into the research activities around children with congenital craniofacial deformities in Sweden.
2) To get involved in the care of children with craniofacial anomalies, several of them born with a syndrome.
3) To learn about the step-wise treatment of children with craniofacial deformities and the overall support to them and their families in a Swedish perspective.
4) To reflect on how quality of life may be affected in this patient group.

Overall organisation: Craniofacial surgery is a subspecialty of plastic surgery and includes treatment of congenital malformations, craniofacial tumors and trauma. The congenital malformations are primarily premature closure of cranial sutures (craniosynostosis) and craniofacial deformities such as Crouzon, Apert, Saethre-Chotzen and Pfeiffers syndrome. Many other syndromes or anomalies are treated like Treacher-Collins syndrome, Binder syndrome, hemifacial microsomia, facial clefts and frontonasal dysplasia. Benign and malignant tumors as well as congenital growths are treated like neurofibromatosis, fibrous dysplasia, dermoid cysts and vascular malformations. Both primary and secondary craniofacial trauma patients are handled. The unit I visited in Gothenburg is a national centre for craniofacial surgery.

The multidisciplinary craniofacial team involves Plastic surgeons, neurosurgeons, anesthetists, oral and maxillofacial surgeons, orthodontists, a psychologist and a speech therapist are all part of the team. The craniofacial unit in Gothenburg performs about 150 intracranial operations a year and follows the patients thoroughly often up to adulthood and sometimes thereafter. There are about 10 consultants at the plastic surgery department, but not all work full time since they have commitments at private clinics and only thre of them work in the craniofacial team. The number of ST/registrars and SHO´s varies.

I arrived at the department of Plastic surgery in Gotheburg on Monday the 16´th of April. There was a meeting at 8 am where acute cases that had arrived during the weekend were reported by the on call team (post take) and the coming weeks operations were discussed. The two consultants in the craniofacial team went thereafter to quickly see patients with their relatives that were planned for operation that day and then to the ward if there were any discharge of patients to do after the weekend. For the moment, no ST/registrars were connected to this highly specialized team but instead two Italian guest registrars (in plastic surgery) that spent 6 months in Gothenburg to learn more about cranial surgery in children.

The weekly timetable involved placement at the theatre Monday, Tuesday and some Wednesdays. There were clinics on Wednesdays and Thursdays and Fridays were usually free after the ward round.

The first case I assisted on was a 6 months old boy with sagittal synostosis which means premature fusion of the sagittal suture, leading to a head shape often referred to as scaphocephalic (boat shaped) and early intervention is necessary for normal development of the brain. After a coronal, zigzag (otherwise the hair will part along the scar) incision is made from ear to ear for access to the frontal and temporal bones. After the osteotomy lines were marked a neurosurgeon came in to do the osteotomy with a special saw. The bone was taken out and further segmented and stripped while the neurosurgeon checked for any bleed or CSF-leakage. The bony parts were then repositioned and springs were installed to prevent new fusion of the bones. The technique with stainless steel springs was introduced in Gothenburg some years ago and the first report on the initial 4 cases was published in 1998 by Lautitzen et al. It is now standard technique to prevent relapse and fusion of bones during growth and the interest from other units is huge with constant visits from all over the world. The springs are usually left for a couple of years before they are removed during a quick procedure. About 50% of the children need a second operation when they are between 5-7 years old.

Some of these children also have facial deformities and/or cleft lip and palate so they are also under care of orthodontists, maxillofacial surgeons and speech therapists. Even if some of the treatment, like correction of a cleft lip and closure of the soft palate, is done well before the child starts school, there might be problems with low self-esteem and even bullying and that is why a psychologist is connected to the team. The final facial correction (jaws, nose etc) is usually done after the growth spurt. A memorable case was a 3 year old boy who had a tracheostomy due to micrognathia and a too small retroglossal space. Using distraction osteogenesis, the mandible was slowly advanced and he could finally get rid of the tracheostomy and breath through the mouth.

Epidemiology: As mentioned above, approximately 150 operations are done every year at the unit. In Sweden, there are about 100.000 live births per year. Apert´s syndrome, which is an autosomal dominant inherited mutation on gene *FGFR-2* in Chromosome 10, occurs in 1 out of 100.000 live births in Sweden and worldwide. This means that only one child per year in Sweden will be born with the syndrome. The same frequency is seen for eg. Pfeifer´s and Nager´s , both autosomal dominant inherited syndromes. Treacher Collins syndrome, a mutation on the *TCOF-1* gene on chromosome 5 is seen in 2 out of 100.000 live births while Goldenhars syndrome is more common and found in 20 out of 100.000 live births in Sweden and internationally. Much more common are the cranial synostosis (premature fusion of the skull bones) which is seen in 1 out of 2000 live births. So there are isolated cranial synostosis and synostosis as a part of the syndromes mentioned above. About 15% of the children with cranial syostosis, also have one of the syndromes mentioned above.

So, if the incidence seems to be the same worldwide, what about care? In countries with a relatively small population, like Sweden, national centres like the one I visited seems reasonable since these children needs very specialized treatment and the surgeons need enough numbers to get experience. In developing countries with large population, treatment is sometimes spread to more units and is probably less organized. Members of the team sometimes visit developing countries in Africa and South America to join other European colleagues to help with surgeries and to organize better care for children with clefts or syndromes affecting the skull or face (eg. Smile train).

In all, this was very interesting placement that gave me a lot of new insight into craniofacial surgery. The whole team was very friendly and professional.