ELECTIVE (SSC5b) REPORT (1200 words)

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

Describe the pattern of congenital craniofacial syndromes in Canada in comparison to other well developed countries (eg UK) and lesser developed countries (eg Vietnam)

Oral clefts are among the most widely known and common craniofacial anomaly, occurring in approximately 1 in every 700 live births worldwide. Oral clefts are often a feature of a syndrome with only 70% of cases being considered non-syndromic. Clefts occur proportionately more commonly in ethnic Asian populations than in ethnic African populations 1. Craniofacial anomalies other than oral clefts occur in 1 in every 1600 new-borns in the USA.

A study on Cleft lips with or without palates (CL/P) and cleft palates without cleft lips (CP) based on information collected between 1992-1998 by 57 international registries (14 from the Americas, 5 from Asia, 2 from Oceania, 36 from Europe) looked at the rates of CL/P and CP, isolated or associated with other defects from a total of 16,923,870 births 2.

In this study, CP had a prevalence range at birth of 1.3-25.3 per 10,000 births, with an overall rate of 5 per 10,000. Rates varied significantly with a low of 2.2 and a high of 8.1 in the different registries. Cleft Palates have the highest report rate in Finland (10 – 14 per 10,000) followed closely by Scotland (8 per 10,000) and Canada. The rates of Cuba, Columbia and South Africa showing the lowest.

In this study, CL/P had a prevalence range at birth between 3.4-22.9 per 10,000 births. The overall rate was 7.9 per 10,000 births. Higher rates were seen in Asian (China and Japan) and South American (Bolivia, Paraguay) registries. The lowest rates are seen in South African and Southern European registries. Cleft lips, with or without palate, is commonest in Bolivia. The highest reported prevalence rate (2.28 per 10,000) was reported in the city of La Paz. This is due to environmental factors, the high altitude and associated chronic hypobaric hypoxia, and genetic factors, the population is of a Mongolic Amerindian ethnicity. Similar high rates of Cleft lips, with or without palate are seen in the ethnic Mongolian population of Tibet who also live at an almost equally high altitude as the Bolivians in La Paz 3-4.

In conclusion, common craniofacial malformations such as CL/P and CP vary wildly with ethnicity and environment. There seems to be less association between rates and the economic development of a nation.

References

1) WHO Human Genetics Programme; Global strategies to reduce the health-care burden of craniofacial anomalies: report of WHO meetings on international collaborative research on craniofacial anomalies, World Health Organization, Geneva (2002)

2) P Mossey, E Castillia; Global registry and database on craniofacial anomalies, World Health Organization, Geneva (2003)

3) Rosano A, Mastroiacovo P (2001) Global distribution of craniofacial anomalies. (Unpublisheddocument presented at the WHO Registry Meeting on Craniofacial Anomalies, Baurú,

Brazil, 4-6 December 2001; available on request from Human Genetics Programme of the World Health Organization, 1211 Geneva 27, Switzerland.)

4) Mossey PA, Little J (2002) Epidemiology of oral clefts: an international perspective. Chapter 12 in Wyszynski DF, ed. Cleft lip and palate. From Origin to Treatment. ISBN: 0-19-513906-2. Oxford, Oxford University Press, 127-158.

Describe the levels of care and treatment available to patients with congenital craniofacial syndromes in Canada compared to other well developed contries (eg UK) and lesser developed countries (eg Vietnam)

Levels of care available in the UK and Canada are identical. Both healthcare systems are well funded and have a layered approach to healthcare. The lowest level of care is primary care. In the UK this level is performed by a general practitioner responsible for caring for the whole family. In Canada primary care is undertaken by a Family Doctor, a doctor responsible for the adults in the family. There is also a community paediatrician who is responsible for the children in the family. This differs from the UK where paediatrics is a secondary care speciality. The next levels of health care in the UK and Canada is secondary and tertiary care. These are the same in both countries and comprise of specialists care. Both seconday and tertiary centres provide intensive care in the UK and Canada.

Therefore a child born with congenital craniofacial malformation has several routes of accessing care in both the UK and Canada. If diagnosed antenatally, the birth can be planned and early consultation with appropriate specialist secondary and tertiary care can occur. This would allow for the child to be worked up and prepped for any potential surgeries that need to occur soonafter birth. If the malformation is diagnosed afterbirth at a non-specialist centre or even in primary care, the child can be easily reffered to a tertiary centre for care of their craniofacial malformation.

In lesser developed countries the access to health care is not as widespread and have many more layers to their healthcare. For example in Kenya the lowest level of care is provided by dispensaries, which are run by nurses and are desgined to treat simple ailments such as the common cold and flu. The next level up is the health centre which has at least one doctor. The health centre's aim is mainly preventative medicine rather than curative focusing mainly on childhood vaccinaitons. A health centre does not come equipped with theatres. The next level up is the sub-district hospital which is similar to the health centre but is managed mainly by doctors and has theatres capable of basic procedures such as caeserian sections. Next is the district hospitals which are the refferal point for sub-district hospitals, these usually have the resources to provide comprehensive medical and surgical services. The next level up is the county hospitals. Kenya has 47 counties each served by a county hospital which is the refferal point for district hospitals. District hospitals are well funded with access to specialist care and intensive care.

The above description of the healthcare system in Kenya shows a potentially lengthy refferal route for a child born with a congenital craniofacial malformation. Only when reaching near the top tier of care in Kenya is intensive care support available making Kenya's ceiling of care equivalent to the UK and Canada's average secondary care.

What public health intiatives exist regarding congenital craniofacial syndromes (and congenital syndromes in general) in Canada and how do these compare to the initiatives that exist in the UK

Antenatal care in Canada and the UK are similar. Both consist of regular antenatal appointments where the well being of the mother and child are checked. Both countries provide blood tests for the screening of common genetic conditions (eg Trisomy 21 and Trisomy 18) and ultrasound scanning for structural abnormalities. Both countries also provide definitve diagnosis via chorionic villus sampling or amniocentisis for at risk mothers.

In the UK a detailed structural scan, known as the mid-pregnancy anomaly scan takes place between 18-21 weeks. This scan would aim to diagnose all sturctural malformations including congenital craniofacial malformations. In Canada a detailed ultrasound scan is offered to all women at 18-20 weeks of gestation.

In families with known syndromic craniofacial malformation genetic testing would be offered and ante-natal diagnosis via chorionic villus sampling or amniocentisis for all subsequent pregnancies. This is available in both the UK and Canada.

Gain exposure to paediatric general surgery and become more comfortable with the pre and post op treatment of a paediatric patient.

The pre-operative assessment of a paediatric patient is similar to that of an adult patient. All patients need baseline bloods including a full blood count, renal function and liver function. Every patient also need a group and save (type and screen) or a cross-match in emergencies. All patient also require an electrocardiogram. Further tests would be determined by the comorbidities of a patient (eg Echocardiogram in a child with congenital heart malformations or in an adult with valvular disease).

Post operatively care differs slightly, especially in regards to feeding. In adults, a patient can be encouraged to feed and a doctor can assume that the patient will eat or drink provided they can tolerate it. In paediatric patients, especially in very young children, feeds have to be estimated and the patient has to be assessed for signs of intolerance (eg reflux, vomiting). Bowel habits are easier to judge in smaller children too.