

PREFACE

My initial interest in childhood blindness was triggered by the number of blind and visually handicapped children I encountered at SJOH in Jerusalem soon after commencing work in 1985. Data on the size of the problem in this region, and from its neighbouring countries, was absent at the time. The unique geopolitical and historical characteristics of each of the WB and GS with its physical separation geographically after 1948 and the interrupted and constrained links between the two parts after 1976, together with the social, economic and anthropological differences of the population could have left an imprint on the pattern of eye disease in each of the regions differently. After 1948, GS became under Egyptian administration and followed the economical sphere of that country, with open route and free travel to any part of Egypt together with the closer social and economic ties between the two that has always existed historically. The educational system and school curriculum was also that of Egypt. The WB on the other hand became an integrated part of the Kingdom of Jordan.

In addition to this, the demographic characteristics of the population in question, the proximity of towns and villages to each other, especially in the GS, made full access to patients and their families feasible which, when combined by the well established custom of the oral tradition of transmitting family history and genealogy throughout the generations, allowed large scale genealogical studies to be made of the recruited patients and families.

This work has focused on 3 aspects of childhood blindness; the first on the epidemiology of visual disability and its causations, the second on the degree of inbreeding and consanguinity on the visual disability and the third on the clinical aspects of the commonest conditions encountered (namely retinal dystrophies, in particular cone and rods disorders which were too frequently encountered in the out patient clinics).

The body of thesis, therefore, is divided into two main chapters; one for the review of the literature on the epidemiology of blindness and consanguinity and a second for the results and analysis of the data. This is in addition

to 4 introductory sections on the history of blindness and SJOH, the history and geography of Palestine and a section on the demography of the Arab world with emphasis on the Palestinians. The purely clinical data on retinal dystrophies and other major conditions in particular CG and microphthalmia has been left out of this dissertation to reduce the bulk of the work and avoid exceeding university guidelines was put aside for future publications.

The first chapter (chapter 2) comprises 7 sections of literature search on blindness, from the earliest attempts to address the condition and collecting data on the blind to the most recent prevention plans and projects. It encompasses a review of the avoidable blinding conditions and blindness worldwide both in adults and children with special emphasis on the Arab world.

The chapter on the 'results' contains 8 sections on the Epidemiological findings in the study starting with evaluating the patients, sibships and pedigrees in the series, visual acuities and classification of the conditions both anatomically and aetiologically in addition to separate sections on non-hereditary conditions and the epidemiology of the common clinical conditions encountered in the study. Marriage patterns and inheritance modes are addressed in a separate section and lastly a section is included containing the various figures extracted from family histories and pedigree charts on prenatal and postnatal mortalities.

The data was initially compiled on special protocol forms and later transferred laboriously into a database in the UK. This was later adopted in line with modified WHO criteria on visual acuities and anatomical and aetiological classifications of conditions ^{11, 12}.

The large bulk of data gathered in the course of this study, the desire to preserve the data and due to the efforts and the collective hard work and dedication in completing the fieldwork coupled with an increasingly demanding NHS clinical commitment upon returning to the UK delayed the finalisation of the work considerably. Despite this, several aspects of the work has already been published and used in lectures and talks in addition to some very valuable molecular genetic work that has been carried out from the blood samples collected (see appendices).

I believe that the length of time taken to accomplish the thesis has not in any way affected the validity, or the significance of the data, particularly given its in-depth detail and the static demographic situation in the area involved. Consanguineous marriages are still commonly practised and remain the preferred choice for a sizeable number of people in this region ⁷¹⁵, consolidated, in the target community, by external factors imposed on these communities that render alternative options unfeasible. The socio-economic conditions have remained poor and, if anything, have worsened as a consequence of the ongoing unrest.

All the work coming from the Arab world, and some Muslim countries, points to the predominance of consanguinity in the regions and the high prevalence of hereditary conditions, but lacks the detailed genealogy and clinical data found in this study.

One possible difference that might have emerged which could have influence the pattern of blindness is the increase in the number of trauma cases resulting from war injuries. This increased after the first uprising ' the Intifadha' and has continued to increase with the ongoing unrest causing considerable decline in healthcare and increase in medical conditions that are associated with increased poverty and hardship ⁷¹⁰.

It is greatly hoped that this work will help to shed light on a forgotten and neglected problem of childhood blindness in these countries and serve to highlight the importance of addressing this vital issue.