

## 13 PATIENTS AND METHODS

### 13.1 Definitions of Childhood

Childhood is defined as 0-15 years inclusive. The WHO categories of visual impairment (vi) define blindness (BL) as a corrected visual acuity in the better eye of less than 3/60, and severe visual impairment (SVI) as corrected visual acuity in the better eye of less than 6/60 but equal to, or better than 3/60.

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### 13.2 Definitions of Childhood Blindness (CB)

Childhood blindness refers to a group of diseases and conditions occurring in childhood or early adolescence, which, if left untreated, result in blindness or severe visual impairment that are likely to be untreatable later in life.<sup>36, 37</sup>

### 13.1 Patients And Schools (Table 13-1)

#### 13.1.1 Introduction

Pupils, trainees and residents in the blind schools, training centres and residential homes in the West Bank (WB) and Gaza Strip (GS) were examined between October 1985 and September 1987 using the Mobile Eye Unit of SJOH (Outreach Programme). The criteria for inclusion in the study were childhood onset bilateral visual impairment in the population of the WB and GS.

#### 13.1.2 Place Of Examination (Plates 15, 16, and 17)

Blind institutes in the WB comprised 3 primary schools for girls funded by local and international charities and 1 school for boys which provided both primary and secondary and was funded by the Department of Education. The former 3 also accommodated very young boys. The remaining 2 institutions were residential for all disabilities and housed older students in higher education. The primary school in Nablus also contained a workshop for older girls who did not proceed to higher education. All the schools in the WB followed the

Jordanian curriculum. Boys progressing to university education who lived away from the universities sites either stayed at the residential homes in the WB or moved to Jordan. A few children from affluent families were sent to Europe and America for further education.

In the GS, there was one mixed primary school funded by UNRWA which also contained a workshop for older girls. The school was non-residential and children who progressed to secondary education joined the sighted school in the GS. Students from Gaza who joined WB universities resided in residential homes in the WB. The schools in the GS followed the Egyptian curriculum.

Each of the blind institutions was visited by the Blind School Survey (BSS) Team using the 'Outreach' Mobile Eye Unit facility of SJOH. The schools' locations and patient numbers are given in Table 13-1 (Plates 18 - 20).

| Institute/Residence                            | Pupils <sup>a</sup>    | Residents | W/L <sup>b</sup> | Relatives  | Total      | Type of Institute           |
|--|------------------------|-----------|------------------|------------|------------|-----------------------------|
| <b>West Bank</b>                               |                        |           |                  |            |            |                             |
| Al-Ala'iyeh (Bethlehem)                        | 46                     | -         | -                | 13         | 59         | Boys, primary and secondary |
| Al-Bireh (Ramalla)                             | 57                     | 7         | 4                | 24         | 92         | Girls, primary, boarding    |
| Al-Shurooq (Beit Hanina)                       | 30                     | 20        | -                | 7          | 57         | Girls primary, boarding     |
| House of Hope (Beit Jala)                      | -                      | 28        | -                | 2          | 30         | Residential home            |
| House of Light (Beit Jala)                     | -                      | 1         | -                | 0          | 1          | Residential home            |
| Al Nour (Nablus) School                        | 5                      | -         | -                | 6          | 11         | Small girls primary         |
| Training Centre                                | 9                      | -         | -                | -          | 9          | Training workshop           |
| <b>Gaza Strip</b>                              |                        |           |                  |            |            |                             |
| UNRWA (Gaza City) School                       | 77                     | -         | 21               | 91         | 189        | Mixed primary               |
| Training workshop                              | 14                     | -         | -                | -          | 14         |                             |
| Residential/multi handicaps home               | 4                      | -         | -                | -          | 4          | Residential/multiple        |
| <b>Sub-Total recruits via schools</b>          | <b>242<sup>c</sup></b> | <b>56</b> | <b>25</b>        | <b>143</b> | <b>466</b> |                             |
| Out-patients (OP) and home visits <sup>c</sup> | 202                    | -         | -                | 1          | 203        | All age groups              |
| <b>Total analysed<sup>d</sup></b>              |                        |           |                  |            | <b>669</b> |                             |

<sup>a</sup> includes pre-school inhabitants and 6 in higher education elsewhere. <sup>b</sup> Pre school children on the schools' waiting list. <sup>c</sup> Also includes relatives and some 85 cases from the OP case notes. <sup>d</sup> This is derived after excluding 40 cases for not fulfilling criteria: 40 OPs and 14 seen at Gaza school.

**Table 13-1: Source of patients in the study**

When circumstances prevented families from attending the school/institute or the hospital, or extensive work was required, (particularly where several members of the family were involved), the team visited the family in their home (Plate 9). This approach was used extensively when collecting genealogy information from the family elders, and relied on the oral tradition. This work was often done with the collaboration of the local social services workers and the village "Mukhtar" (village chief of mayor of a village).

### **13.1.3 Patients**

The number of cases initially enrolled was 709, 40 of them were later excluded from the study for not fulfilling the study criteria of childhood onset bilateral visual impairment in the WB and GS, leaving 699 cases. Table 13-1 gives a breakdown of the patients, their source and their schools. All the pupils, trainees, blind teachers, their affected siblings and younger children on the school's waiting lists were examined. Outpatients meeting the criteria were also included in the study. Parents and siblings of the affected patients were also screened.

## **13.2 The Team**

The team included a dedicated Blind Schools Survey team and the Outreach Programme team. The BSS team was comprised of a full-time research assistant/social worker (from June 1986); one part-time personal secretary/research assistant (from October 1986); and one full time junior research assistant who was later trained as an electrodiagnostic assistant technician (from March 1987). The Outreach team comprised a driver/paramedic; 1 permanent outreach nurse; and an additional 1 to 2 non-permanent nurses. The visiting orthoptist joined the team when time permitted; alternatively patients were sent to SJOH for orthoptic assessment.

## **13.3 Protocol**

### **13.3.1 Protocol Forms** (Appendices 2, 3, 4)

Forms for family history, clinical examination, and sibship data were designed using an Amstrad word processor computer. A special classification and numbering system (cross-indexed) was designed for the study. An instru-

ction booklet<sup>712</sup> was prepared for the Outreach Team and included, the research protocol, instruments required, medications etc.

### **13.3.2 Questionnaires**

Questionnaires were devised in Arabic to assist the team and the resident social workers to obtain information from the parents about their siblings prior to the home visit and this information was then transferred to the equivalent questionnaires in English by the trained bi-lingual social worker.

Specially prepared protocol forms were designed for family history and sibling information including prenatal, perinatal, neonatal, postnatal and adolescent history.

### **13.3.3 Natal Histories**

Prenatal, perinatal and postnatal data together with full ophthalmic, medical and genetic histories were taken from the patients and/or their parents. This included the onset of the first symptoms, the presenting symptoms or features and the time of their onset, the course of the disease and the presence and/or absence of photophobia and night blindness. Data on the extended pedigrees, helped by the strong oral tradition in this community, was taken from older patients, their parents and/or accompanying relatives. Detailed pedigree charts were drawn from the compiled data.

All available data from the SJOH medical records and regional social services departments were obtained.

### **13.3.4 Demographic Data**

We obtained personal details of the patients, their siblings and half siblings where possible and their parents, including details of the mother's family and the extended family "Hamoula".

In addition, we ascertained whether pedigrees were from the same location (village/town), whether they had refugee status, and whether they lived in or outside a camp. Information was also obtained on the origins of the family, ie their origin and location pre and post 1948 and pre and post 1967, and their present location and residence. This included information on whether spouses were from the same locality originally, particularly as a proportion of the patients

were refugees and came from villages or towns which no longer existed or were now contained within the State of Israel.

#### **13.3.5 Genetic Protocol**

Full marriage details were asked including the degree of consanguinity as far back as was known accurately (Classification according to Curt Stern 1973). Pedigree charts were drawn from the data available and frequently checked and revised to eradicate errors, especially in the large pedigrees. Patients were placed on the chart and other family members were plotted. The exact sequence and gender of births, including any prenatal death was observed with as much accuracy as possible. The degree and extent of cousin marriage was recorded and traced as far back as possible on the pedigree chart. Where it was certain that marriage was from the same family, but the exact relationship could not be ascertained with certainty, either from history or on the family chart, it was recorded as 'same family'. If consanguinity was absent, the geographic origin of the parents was investigated ie whether they came the same village, town or district etc. 'No relationship' was marked on the family chart when this was stated and confirmed.

#### **13.3.6 Ophthalmic Examination**

Full ophthalmic examination included visual acuities, cycloplegic refraction, orthoptic examination, slit-lamp biomicroscopy and fundoscopy with mydriasis was performed the Author. Visual field assessment was carried out by confrontation method but, when possible, Goldman perimetry was performed at the base hospital.

Apart from the cases where information was extracted from SJOH casenotes at the very end of the study, all patients were examined by the author. When necessary patients were referred to Dr Lee Chumbley, then, Consultant and Director of Research and frequently referred to weekly staff combined clinical meeting. Additional ocular examination of some selected conditions was also conducted by Mr Anthony Moore, Consultant Ophthalmologist, at Addenbrooke's Hospital, Cambridge, who was invited to offer his advice and accompanied the author and the Outreach Team for this purpose.

When further assessment was required, e.g. examination under anaesthesia, fundus photography and fundus fluorescein angiography or orthoptic follow-up, patients were seen at the base hospital. Electrodiagnostic tests were performed three months prior to the end of the survey when facilities became available.

Dental work was initially carried out by a local volunteer dentist who participated in a one-day fieldwork with the author. Later, Dr David Smith, Professor of Dental Radiology, Kings College Dental School, London, became involved and he visited the WB and GS and screened all patients with any suggested dental abnormality and also the relatives of patients.

Patients who required medical opinion were referred to either St Joseph's Hospital (Neurology and ENT) or Al-Makasid Hospital for medical conditions. Surgical opinion was provided for this research on the SJOH site by Dr Peter Qumri, Consultant Surgeon at King Al-Hussein Hospital, Beit Jala.

### **13.3.7 Assessment of Visual Functions**

Visual acuities, both unaided and best corrected, were measured using Sheridan Gardiner test charts and, for the very young, cake decorations. When necessary, E-test chart was used.

Colour vision testing was performed, when indicated, using Ishihara Pseudo-isochromatic Plates, City University plates, text reference for colour to assess gross colour and in the severely visually impaired, Kodak red filter C 25, and the green and blue filters of the slit lamp.

Visual fields testing was by the confrontation method and when necessary with portable Bjerrum screen. Goldman Perimetry was used at SJOH when possible.

Electrodiagnostic assessment, including the setting up of normal values and training, was carried out by Mr Chris Hogg, Senior Technician, the Electrodiagnostic Unit, Moorfields Eye Hospital, London.

## 13.4 Data Analysis and Parameters Used

### 13.4.1 Software

Data was compiled on the special protocol forms and later transferred to a database (Microsoft Access). A spreadsheet (Microsoft Excel) was used for calculation, Tables and graphs.

### 13.4.2 Visual Acutities

Visual acuity levels were analysed according to WHO/PBL categories (WHO/PBL Eye Examination Record for Children with Blindness and Low Vision, Coding Instruction) <sup>11, 12</sup>. The same protocol was applied for anatomical and clinical classification including where there was a difference in the anatomical diagnoses between each eye in which case the diagnosis of the Best Seeing Eye was used. For the visual acutities, WHO categories for visual acutities were used in the analysis. (Table 13-2).

| <b>V A Category</b>   | <b>Description</b>  | <b>Visual Acutities</b> |
|---|---|-------------------------|
| Category '1'  | No impairment   | 6/18 or better          |
| Category '2'  | Visual impairment   | <6/18 to 6/60           |
| Category '3'  | Severe visual impairment  | <6/60 to 3/60           |
| Category '4'  | Blindness   | <3/60 to LP             |
| Category '5'  | Blindness   | NLP                     |
| <i>Patients on whom reliable assessment of vision was not possible have been categorised by the WHO under categories '6' and '7'.<br/>Unavailable data was included under a new category 8.</i> |   |                         |
| Category '6'  | Believed to be sighted but with some visual impairment              |                         |
| Category '7'  | Believed to have significant visual disability or to be blind.      |                         |
| Category '8'  | No reading is available, such as unable to examine, lost data, etc. |                         |

**Table 13-2: WHO categories of visual acutities**

### **13.4.3 Classification Methods**

Conditions were initially classified clinically but later, the WHO modified categories on anatomical and aetiological classifications were applied <sup>11</sup>.

## **13.5 Additional Protocols**

Two additional elements were later added to the protocol. These were:

### **13.5.1 IQ and skills** (Tables 22-18, 22-19)

The level of intelligence and any special skills judged by the degree of the patients' educational Performance using school examination marks as a parameter in the various subjects was evaluated. Randomly selected samples in proportion to the size of cohort in each condition was planned. This was not possible to achieve fully as a bias of the schools towards their better achievers resulted in under representation of some conditions.

### **13.5.2 Red-Tinted Glasses** (Plate 54)

The Efficacy Of Red-Tinted Glasses in providing symptomatic relief and alleviating photophobia to photophobic patients and comparing them to commercially available types of tinted sun glasses.

## **13.6 Reports to Social Services**

Following each examination, medical reports were prepared on the pupils and were provided for the local social services indicating the level of the visual acuities and the visual disability together with any other relevant information. The same format was later officially adopted by the hospital and the Social Services Department.