TREATMENT PROFILE FOR CLEFT LIP AND PALATE

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A thesis submitted in partial fulfilment
for the degree
MASTERS IN DENTAL SURGERY
(PAEDIATRIC DENTISTRY)

DEPARTMENT OF DENTISTRY
FACULTY OF DENTISTRY
THE UNIVERSITY OF ADELAIDE

1995

Errata

p. 13,	line	7	occurring in conjunction ...
p. 13,	line	9	Robin sequence, ~~Syndrome, Crouzons disease~~ Hemifacial
p. 16,	line	10	Unit ~~since~~ during the last 25 years
p. 16,	line	14	considers that the
p. 16,	line	16	Sex predominantce
p. 16,	line	17	~~O~~other
p. 16,	line	18	~~with~~ to pregnancy
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p. 25,	line	3	~~down's~~ Down syndrome ~~spinal~~ spina bifida
p. 25,	line	11	into_seasonal.
p. 25,	line	16	to research a significant
p. 29,	line	6	situs inverseus
p. 31,	line	15	who have an
p. 32,	line	14	hypotheses ~~and~~ making use of
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p. 38,	line	6	severe myopia
p. 39,	line	9	But, an assessment of the
p. 40,	line	14	infants is over
p. 40,	line	17	has been the most
p. 44,	line	11	treatment ~~are~~ is generally
p. 45,	line	7	after ~~and~~ full orthopaedic
p. 47,	line	12	The secondary reconstruction of cleft
p. 52,	line	8	Craniofacial
p. 55,	line	5	successfully. ~~Their~~ There is also
p. 58,	line	5	the ~~tetragens~~ teratogens that are
p. 58,	line	14	studies are ~~requiered~~ required to analyse

p. 58,	line	15	Cranio-~~F~~facial
p. 59,	line	3	This will also enable
p. 59,	line	6	aimed ~~in~~ to provide
p. 59,	line	14	team, ~~and~~ the patient
p. 59,	line	18	~~bring about~~ enable more comprehensive, coordinated care to be provided
p. 61,	line	9	a cleft requires
p. 61,	lines	11, 12	which ~~comprises~~ consists ~~of staff of~~ an interdisciplinary team, ~~and~~ all the team members of which are trained and
p. 62,	line	10	between 1940 ~~to~~ and 1993
p. 62,	line	11	the subject had a cleft of the lip
p. 62,	line	13	all the subjects selected
p. 63,	line	8	deceased ~~after~~ a few months after ~~of~~ birth
p. 63,	lines	17, 18, 19	for permission ~~of~~ to conduct the study. ~~and at~~ At the same time permission to assess the patients case notes for the study was requested from the Medical Records Department and Cranio-~~F~~facial Unit. ~~Permission to assess the patients case notes for the study was requested from them.~~ The
p. 64,	line	8	The Medical Records Department of the Adelaide~~'s~~
p. 64,	line	11	helped in ~~removal~~ retrieval of the
p. 64,	line	15	CFU1 form. ~~Prepared.~~
p. 69,	line	8	Harw~~v~~ard
p. 75,	line	13	interventions
p. 75,	line	15	normal ~~occlusion~~ occlusal development.
p. 75,	line	16	Any ~~any~~ small
p. 75,	line	16	long way ~~in~~ towards
p. 75,	line	17	employed in the
p. 75,	line	18	palate as a whole
p. 76,	line	3	according to the following cohorts, ~~defined as~~
p. 80,	line	20	to quite a low
p. 80,	line	21	value for
p. 88,	line	7	for the 1990s

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p. 93,	line	14	age of 12 years, Ffollowed
p. 100,	line	6	ranged ~~to~~ from a minimum of six days to a maximum of 50
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p. 111,	line	7	value ~~were~~ was
p. 111,	line	8	The standard deviation
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p. 115,	line	5	visit ~~with~~ and counselling
p. 115,	line	6	feeding ~~is done by~~ from the
p. 116,	line	3	age, nasendoscopy is ~~done~~ undertaken.
p. 116,	line	13	find ~~weather~~ whether the patient category of needing orthodontics alone or whether they
p. 116,	line	14	requires
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p. 117,	line	6	meetings ~~along with~~ which include the
p. 117,	line	7	basis, after the annual assessment
p. 117,	line	8	development ~~with~~ by the orthodontist. ~~And~~ These meetings
p. 117,	line	9	the parents, ~~consisting of~~ and provide the family with a complete
p. 117,	line	16	any ~~touch up or~~ secondary surgery ~~are~~ is required
p. 118,	line	1	time the patient reaches

p. 119,	line	3	255 cases ~~were~~ was only
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p. 136	CHARLTON	malformations in
p. 136	COHEN	naosologic
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p. 139	HALL	Hall ~~m~~Medical
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p. 140	MARAZITA	in ~~s~~Shanghai, China;
p. 141	MILLARD	~~w~~Wilkins
p. 145	WALLACE	Severe
p. 145	WITT	In ~~Robert~~ A Hardesty (edi):

Minor amendments

p. 1 Approximately one child in 600 live births is born with a cleft of lip~~, palate, or both structures~~ and palate and one in 1,000 with a cleft palate alone.

p. 76 The number of cases incorporated in the study was 255. Of these 245 were available for analysis of first interventions by birth cohort. These were distributed as follows:

1940–59	10
1960–69	14
1970–79	82
1980–89	133
1990–93	6
Total	245

SUMMARY

Approximately one child in 600 live births is born with a cleft of lip, palate, or both structures. These deformities have a profound emotional effect both on parents and child and also seriously compromise physical and psychological well being of the child affected with the deformity.

Treatment procedures and regimes for the repair of cleft lip and palate have long been available, but special cleft palate clinics, offering multidisciplinary services, have been established only in last two decades.

The treatment of cleft lip and palate patients requires multidisciplinary cooperation. The spectrum of outcomes of the surgical interventions for the repair is considerable and may be related to the type of surgical technique performed, timing sequence and skill of the individual surgeon.

This study aimed to describe and analyse the multidisciplinary approach in correction of cleft lip and palate at **The Australian Cranio Facial Unit** based at the **Adelaide Children's Hospital**. Case notes of children admitted at the Adelaide Children's Hospital over last 25 years for treatment of cleft lip and palate were examined. This thesis is centred on the types of surgical technique performed

on the child and special emphasis is given to the age of patient at the time of each surgical intervention and thereby analysis of the treatment regime followed.

Different types of cleft classifications are listed. The aetiological background for facial clefts are described. Associated malformations or syndromes are listed. Descriptions are provided for many of the commonly associated syndromes or malformations. Sex predominance, blood grouping, maternal age and problems related to pregnancy are noted.

The results of the study are outlined and graphically represented. However, the main findings obtained from the data showed 48.6% of the study population had combined cleft of anterior and posterior palate (Group II). Female : male ratio was found to be 1:1.71. Mean age of first intervention of various surgical interventions by cohort was seen to be decreasing with a relatively small deviation around the mean in more recent times. The mean age of intervention for speech therapy has decreased considerably, but the standard deviation has increased considerably for the 1990s cohort showing the intervention has been carried out from less than one year to five years of age.

Analysis done to obtain the of age distribution of the study sample for various surgical interventions showed 87.9% of the study sample had repair of cleft lip at the age of 3-4 months. Age of first intervention for repair of cleft palate showed

74.9% of the study subjects had it at the age of 6-12 months. For alveolar bone grafting 32% of the sample had it at the age of 12 years. Pharyngoplasty showed a bimodal distribution: the first peak was at 4 years of age and second at 7 years of age. The distribution for nasal tip revision showed 34.8% of the study subjects had it at 12 years of age. 27.4% of the study sample had their first bilateral myringotomy at the age of 1-5 years and osteotomies were done at an age of 15 years of age for 44% of the study subjects. The complete distribution for the various surgical interventions are described and comparison is made between the various birth cohorts in the study.

The results stress the value of a coordinated treatment plan involving many people and disciplines as illustrated in the operation of Australian Cranio Facial Unit .The results also indicated the worth of objective speech and facial growth evaluation. The limitations of the present study are discussed and continued research into the field of facial clefts is encouraged.

ACKNOWLEDGMENTS

I am deeply indebted to Professor A. John Spencer, Head of Social and Preventive Dentistry, The University of Adelaide, firstly for his kind consent and all his time and assistance which led to the compiling of this thesis. His guidance, patience, suggestions and encouragement were indeed invaluable and made the task of compiling this manuscript an enjoyable and satisfying experience.

A valuable stimulus to my studies was provided by Mr. David J. David, Head of the Cranio Facial Unit, Adelaide Children's Hospital, by way of his assistance in allowing me to conduct the study of treatment profile for cleft lip and palate patients which contributed to the production of this thesis. His willingness to help at every stage was much appreciated.

I am very grateful to Ms Joan Alister, Research officer in the Department of Social and Preventive Dentistry, The University of Adelaide for providing valuable guidance in addition to the computer programmes and analysis of the results for this thesis.

My thanks are due to all the staff at the Medical Records Section, Adelaide Children's Hospital for helping me get access to the patients case notes.

I would have it known, that I am also grateful to many different staff in Social and Preventive Dentistry for giving me assistance and providing facilities whenever required.

Finally, a big thanks to my loved ones and my parents in particular without whose constant support, patience and encouragement things could have been difficult and different.

SIGNED STATEMENT

This thesis is submitted in partial fulfilment of the requirements of the Degree of Master of Dental Surgery in the University of Adelaide.

The thesis contains no material which has been accepted for the award of any other degree or diploma in any University. To the best of my knowledge and belief it contains no material previously published or written by another person except when due reference is made in the text of the report.

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NAME: DR. SONALI SANGHVI COURSE: M.D.S. PEDIATRICS.

I give consent to this copy of my thesis, when deposited in the University Libraries, being available for photocopying and loan.

SIGNATURE: ... DATE: 12/1/96.

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1 INTRODUCTION

Clefts of lip, alveolus, hard and soft palates are the most common congenital defects of the orofacial structures. These cranio facial birth defects affect approximately one in six hundred new born each year in Australia (Hall, 1994).

Approximately one half of the infants affected have associated malformations, either minor or major occurring conjunction with the clefts. However, the incidence figures for more complex anomalies and syndromes such as Pierre Robin syndrome, Cruzons disease, Hemifacial microsomia, etc, are much lower than that for cleft lip and palate(Jones, 1988; Rollnick, 1981).

The impact of cranio facial birth defects is two fold:

(a) that on the patient and the family; and

(b) on society as a whole.

The health and well being of all these children is dependent upon the clinical expertise of those who serve them. In addition, the society as a

whole is affected by the quality of their care because the potential of the affected individual for a positive contribution to the community is inevitably influenced by the adequacy of the treatment.

At present our knowledge of the teratogens that are associated with facial clefting is very limited. It is said that it may have monogenic or polygenic aetiology. However, there are many possible aetiological factors with both genetic and environmental factors contributing to cleft formation (Hall, 1994; Mc Comb, 1989).

At present, the prevention of most clefts is not possible as we yet do not know the precise aetiology. Hence, our main aim is to be able to provide good treatment regime to these children who have special health care needs.

Treatment of clefts involves several fundamental principles regarding the optimal care of the patients:

1. management of these patients is best provided by an interdisciplinary team of specialists;

2. proper diagnosis of the defect and treatment planning;

3. assisting the family in adjusting to the birth of child with a cranio facial anomaly and to the consequent demands and stress placed upon the family;

4. giving information to parents or guardians upon recommended treatment procedures, options and cost to assist them in
   (a) making decisions on child's behalf, and
   (b) preparing the child and themselves for the surgery;

5. arranging meetings with family to initiate family participation and collaboration in treatment planning, later the participation of the grown child in the in treatment decision

6. Monitoring both short term and long term treatment outcomes. Hence, regular follow ups of patients, including appropriate documentation and record keeping is required; and

7. evaluation of treatment outcomes and consideration of patient satisfaction and psychological well being of the patient along with effect on growth, function and appearance.

There is a vast amount of literature on cleft lip and palate treatment today but to date there has been no internationally agreed treatment regime for the various clefts

This study aimed to describe the multidisciplinary approach in correction of cleft lip and palate by Australian Cranio Facial Unit based at the Adelaide Children's Hospital and to analyse:

(a) the treatment regime followed by the Unit since last 25 years

(b) changes of treatment plan over time;

(c) comparisons of various treatment regimes followed at other centres.

It is emphasised that the Australian Cranio Facial Unit considers the multidisciplinary approach is necessary in management of cleft lip and palate. Sex predominant and type of clefts related to sex focused by the data collection is mentioned and Other findings such as blood grouping, family history, and problems related with pregnancy are described

It is hoped that the findings will be useful in analysing treatment procedures and help in making decisions about correct sequence of repair in regard to the effects of muscle balance , scaring and psychological well being of the patient. This may stimulate the development of improved treatment strategies and techniques which would benefit our patients and improve the effectiveness of multidisciplinary treatment.

1.1 CLASSIFICATION

In order to compare dental anomalies associated with various types of clefts, or to develop continuity between diagnosis and treatment modalities, a precise and accurate system of identification is needed. There have been many attempts at classification, but only a few have found wide clinical acceptance.

Clefts of the lip, alveolus and of palate are broadly classified into three groups in the classification proposed by Davis and Ritchie(1922) :

Group I : Prealveolar clefts- unilateral, median, or bilateral;

Group II : Postalveolar clefts involving the soft palate only, the soft and hard palates, or a submucous clefts;

Group III : Alveolar clefts- unilateral, bilateral, or median.

In 1958, Kernahan and Stark suggested a classification based more on embryology than morphology. They proposed three distinct groups:

I : Clefts lying anterior to the incisive foramen, ie, clefts occurring in the "primary palate" as a result of failure of proper mesodermal penetration. This group would include clefts varying from a minor cleft of the lip to those involving the whole premaxilla;

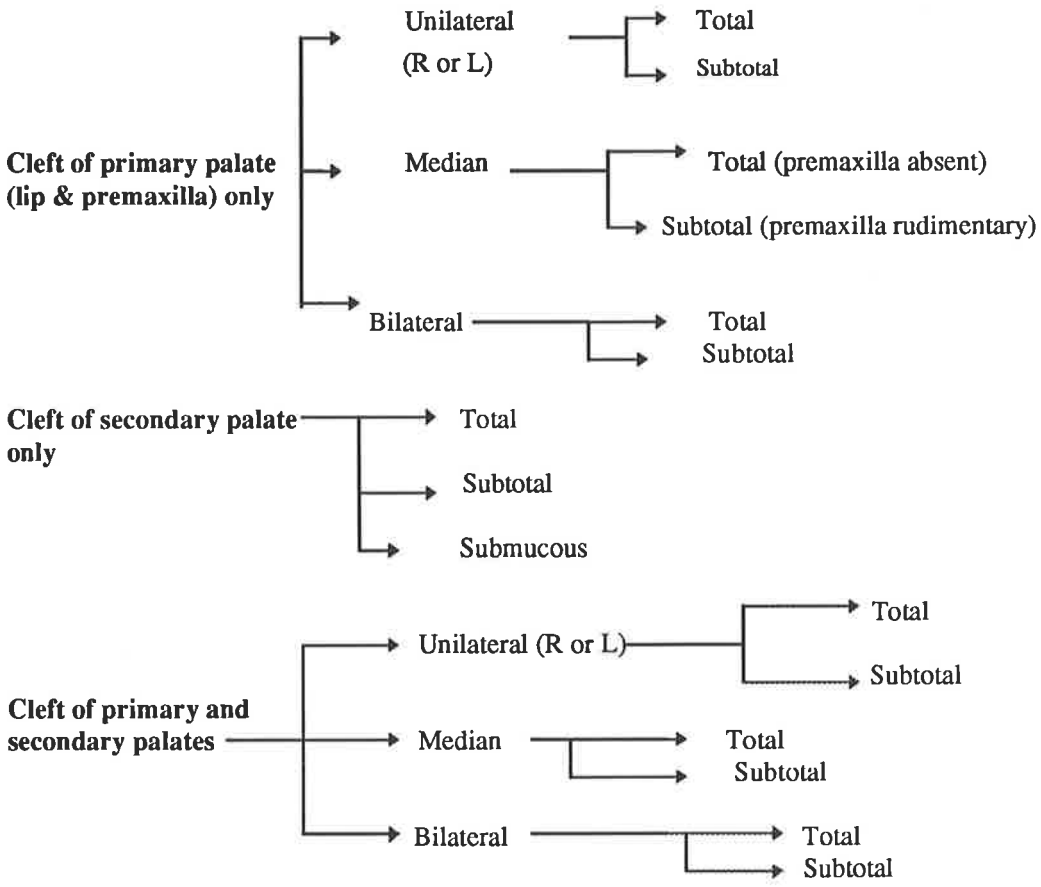
II : Clefts lying posterior to the incisive foramen, ie, those due to a failure of the fusion of the two palatal processes to form the secondary palate;

III : Clefts which combine these two important embryological events, ie, failure of the normal development of both the primary and the secondary palate.

The complete classification is shown diagrammatically in Table 1.1

TABLE -1.1

Revised classification of cleft lip and palate by Kernahan and Stark (1958)



In the above classification it was the incisive foramen and not the alveolus that was considered as the deviding point between the different groups of deformities. In 1971 Kernahan DA proposed another classification the "stippled Y" which proved to be more useful for charting clefts in medical records and describing surgery. This simple "Y" was recently been extended incorporating

Elsahy's 'peaks' to describe nasal floor involvement in partial clefts

(Figure 1.1)(Hall, 1994).

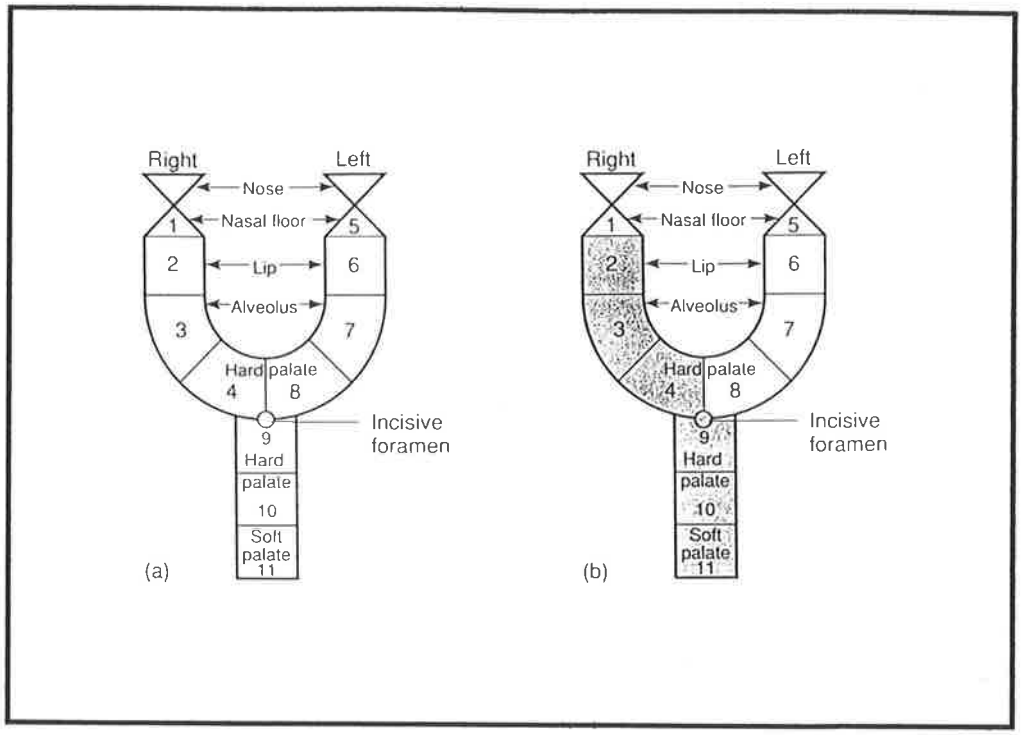


Figure 1.1: **(a)** The 'Stippled Y' classification of clefts of nose, lip and palate ready for shading (from Kernahan, modified by Elsahy cited by Hall, 1994)

(b) The Y-stipple shaded for a Group 2 abcd (R) cleft (right unilateral complete cleft lip and palate with nasal floor involvement and moderate distortion of the nose

In 1987 an international classification of clefts of lip, alveolus and palate was proposed based on the classification suggested at the International Conference of Plastic Surgeons (1967) (Table 1.2) which is ideal for descriptive purposes

and this classification is incorporated for data collection for this study (Hall, 1994)

Table 1.2: International classification of clefts of lip, alveolus and palate (Hall, 1994)

Group 1: Clefts of anterior (primary) palate

a. Lip cleft complete

(a) Lip cleft partial

b. Alveolar cleft including dental lamina

(b) No alveolar bony cleft but dental lamina involvement,
in lateral incisor region beneath lip cleft, as evidenced by the presence of :
conical tooth
(absence of) lateral incisor tooth
fusion/gemination
enamel hypoplasia of tooth crown
other change in tooth morphology
odontome

Group 2 : Clefts of anterior (primary) and posterior (secondary) palate

a. Lip

b. Alveolus

c. Hard palate

d. Soft palate, usually total cleft (if not see below for Group 3)

Group 3 : Cleft of posterior (secondary) palate

c. Hard palate

d. Soft palate, degree of partial involvement designated by one-third, two-thirds or submucous

1.2 AETIOLOGY AND DEVELOPMENTAL PATHOGENESIS

Clefts of the lip, alveolus and palate have been known to be caused by many aetiological factors. Some of them are caused by single mutant genes, some by chromosomal aberrations, some by specific environmental agents and some may be caused by interaction of genetic and environmental factors, sometimes refereed to as "multifactorial."(Fraser 1969; Hall, 1994)

All parents who have a child with cleft are mainly interested in the answer to two questions :

a. Why it happened to their child? Whether they caused it by something they did that they should not have done or something they did not do that they should have done.

b. What are the chances of their having another child with the similar problem?

The first question addresses aetiology. Initially parents are concerned about the amenability of the obvious structural defect to reconstruction; however, concerns regarding the impact of the condition on growth, cognitive function, and social integration are also paramount. Many conditions that cause clefting are associated with other structural or functional problems that have a direct

impact on these areas. Guidance becomes problematic in the absence of an aetiological diagnosis. The second question relates to re-occurrence risk or genetic counselling. Accurate information regarding risk and options for prenatal diagnosis can be provided only if the cause of the problem is clearly identified. Hence dysmorphology and clinical genetic evaluation should be an integral part of patient management. (McComb, 1989; Rollnic et al, 1981).

1.2.1 TYPE OF CLEFT AND SEX

The overall sex ratio of those affected with cleft lip and/or palate is seen to favour males in comparison to females (Moller, 1965; Chi, S 1970; Drillien et al, 1966). The samples of eight large studies were combined by Green in 1963 who calculated a male: female ratio of 58:42 from a total of 3,907 subjects. There appears to be male predominance with the increase in the severity of the deformity, ie, it is greater for cleft lip and palate than for cleft lip (Fogh-Anderson, 1942; Rank and Thomson, 1960) and for bilateral than unilateral defects (Fogh-Anderson, 1942). This is what one would expect on the hypothesis of multifactorial causation, but there are enough exceptions reported (Drillien et al, 1966; Meskin et al, 1968) that the matter cannot be considered settled.

1.2.2 SEASONAL OCCURRENCE

Many diseases have been analysed for seasonal pattern for occurrence. These analyses have been applied also to congenital malformations such as anecephaly, hydrocephaly, down's syndrome, spinal bifida, congenital dislocation of hip and cleft lip and palate (Woolf et al, 1963). This suggests that a seasonal association with incidence would provide evidence of a non-genetic aetiological factor. Wehrung and Hay (1970) suggested that if a seasonal trend is demonstrated for a congenital malformation, concomitant trends can be looked for in other factors such as infectious diseases, ingestion of drugs, malnutrition or availability of certain nutrients, and many other factors.

There are equivocal results of research intoseasonal association with the occurrence of cleft defects. Knox and Braithwaite (1963) from surgical record data and Gilmore and Hofman (1966) from birth certificates reported no association, although both sets of data were not analysed in relation to seasonal variation in normal births.

Woolf et al (1963) recorded the month of birth for a total sample of 889 subjects with clefts. He used Chi- square tests to search a significant deviation in seasonal variation between births with cleft defects and normal births. No significant seasonal trend was demonstrated for cleft lip or cleft palate. On the

other hand Fujino et al (1963) reported on 2,828 cleft cases, using similar statistical methods to those of Woolf et al (1963). Cleft of lip and palate births were seen to be decreased among persons born in Japan during winter(from December to February) and were increased among those born in spring(from March to May). The deviation from random expectation was significant at the one per-cent level in cleft of lip and insignificant, although the trend was similar, for cleft of lip and palate. No seasonal influence could be demonstrated for cleft palate births.

Charlton (1966) reported that for cleft lip and palate cases (136 cases) the Adelaide data showed significant seasonal variation, the highest incidences occurring in June and July. No other type of cleft showed significant seasonal variation in either the Brisbane or Adelaide data.

The establishment of the National Cleft Lip and Palate Intelligence Service enabled Wehrung and Hay (1970) to report from birth certificates on approximately 10,000 cases of cleft lip and palates in the U.S.A.
A systematic sample consisting of approximately 99,000 birth certificates served as a control group. Adjustments were made for the different number of days in each month and the seasonal variation in normal live births. For the total geographical and climatic area under analysis, only cleft lip and palate was significantly different from the controls. The simple harmonic curve fitted to the

data in the Edwards analysis gave the maximal incidence in March. When data were broken down according to climatic area of birth, cleft lip and palate occurrence was seasonally significant only in the hot summer- moderate winter region. The maximal incidence occurred in January which preceded the national trend (March) by about two months (Edwards, 1960).

1.2.3 ENVIRONMENTAL AGENTS

The literature is replete with studies documenting the effects of a variety of physical and chemical agents on cleft lip and palate development. Most of these studies are based on animal experimentation, and it is believed that teratogens are species specific in their effects. Teratogenesis in animals does not imply teratogenesis in humans (Hardesty, 1993).

At present our knowledge of the teratogens that are associated with facial clefting is very limited. Only a few substances such as 13-cis-retinoic acid (used in treatment of acne and psoriasis), alcohol, anticonvulsants have been confirmed as teratogens with direct effects on facial morphogenesis (Gorlin et al, 1990). There are studies going on at present about the association of parental age with incidence of cleft lip and palate and it is believed that there may be a small increase in incidence of cleft lip and palate with increasing parental age (Fraser and Calnan, 1961; Woolf, 1963; Greene et al, 1964; Meskin and Pruzansky, 1968).

Drillien et al (1966) found an increase of threatened abortion and severe vomiting in pregnancies leading to cleft of lip and or palate.

The agent that has been studied most extensively is maternal smoking. Several well designed, case-control studies have suggested between a two to six - fold increase in relative risk for cleft among smokers (Khoury et al, 1987; Khoury et al, 1989). Other equally well designed investigations have yielded negative results (Werler et al, 1990).

1.2.4 FAMILY HISTORY

Familial clustering of cleft lip/plate is consistently found in different populations and supports the hypothesis that genetic factors are important in the pathogenesis of cleft lip/palate (Farrall et al, 1992; Melnick et al, 1980).

This phenotype-genotype association between cleft lip/palate and transforming growth factor alpha locus has been independently confirmed by Chene vix-Trench et al, (1991) in Australian white population and by Holder et al in British white population.

1.2.4.1 *Relationship between Population Frequency and Frequency in Relatives*

Using the threshold model, and making a number of reasonable assumptions, it is possible to derive a relationship between the frequency of the condition in

relatives and the frequency in the general population. If the phenotypic correlation between first degree relatives is 0.5 (that is, heritability is high), the frequency in first degree relatives in proportion to the population frequency approximates the square root of the population frequency (Edwards, 1960). A number of conditions have been shown to fit the criterion for multifactorial causation, including congenital hypertrophic pyloric stenosis, situs inverses (Newcombe, 1963) and atrial septal defect (Nora et al, 1967). For cleft lip/palate, using a population frequency of one per 1,000, this relationship would lead to an expected re-occurrence risk in siblings of 3.2 per-cent, which is well within the observed range (Fogh-Andersen, 1942; Curtis et al, 1961: Woolf et al, 1963).

1.2.4.2 *Twins*

For any condition determined in part by genetic factors, the concordance rate is expected to be higher in monozygotic than in dizygotic pairs. Data on twins with cleft lip and palate, ascertained without respect to concordance are fairly scanty, but do show a higher concordance rate in monozygotic pairs. Twin data are notoriously subject to bias, therefore more research in this field is required (Metrakos et al, 1958; Fraser, 1970; Farrall et al, 1992).

1.2.4.3 *Parental Consanguinity*

The association of rare, recessively inherited diseases with parental consanguinity is well known. Less widely recognised is the fact that the frequency of multifactorially determined conditions can be expected to be elevated in the offspring of consanguineous mating. This results from the fact that consanguinity increases homozygosity, which will increase the proportion of individuals at the tail of the distribution (Newcombe, 1963).

There are very few data available on the offspring of consanguineous matings. The largest available data set on the offspring of consanguineous marriages do not report of any increase in frequency of cleft lip/palate (Schull and Neel, 1965), and nor do any other studies (Sutter and Tabah, 1954; Slatis et al, 1958). Other data collected on parental consanguinity using different approaches also did not give any convincing evidence on occurrence of cleft lip/palate in their offspring(Fogh-Anderson, 1942; Curtis et al, 1961; Fugino et al, 1963).

1.2.4.4 *Other Family Data*

The multifactorial/threshold model makes several predictions, specially:

1. the defect will cluster in the families.
2. the risk for first-degree relatives of affected individuals (parents, siblings, and offspring) will approximate the square root of the population risk.

3. the risk for second-degree relatives (uncles, aunts, half-siblings) will lower the risk for first degree relatives.
4. the more severe the malformation, the greater is the risk of re-occurrence.
5. the greater the number of affected family members, the greater the risk for recurrence.
6. the risk for recurrence will be increased for relatives of the least affected sex, if sex differences are noted.

Recently several investigators reanalysed previously published data sets with respect to a variety of alternative hypotheses, most of which assume the impact of a single major dominantly or recessively inherited gene. Though none of the hypotheses has explained the observed data for cleft lip/palate alone, several reviews involving multiple ethnic groups have supported a major single gene locus effect for cleft lip/palate (Chung et al, 1986; Hecht et al, 1992; Marazita et al, 1992). According to the threshold model, unaffected parents who have affected child carry more than the average number of genes contributing to the condition. Thus the risk of their subsequent children being affected is above average and if their second affected child is born, the parents can be considered to carry still more predisposing genes. The risk for siblings born of unaffected parents increases from 4% after one affected child to 9% after two affected children (Curtis et al, 1961).

If the existence of an affected first degree relative increases the recurrence risk is reasonable to suppose that the same will be true for affected relatives of more distant degree, though the increase would be smaller. No such increase has been demonstrated for cleft lip/palate patients (Curtis et al, 1961).

It has been demonstrated that the frequency of the condition in the near relatives of the patient ought to be higher when the patient is of the sex less often affected. This was first demonstrated for congenital hypertrophic pyloric stenosis (Carter, 1965). This relationship appears to be true also for cleft lip and palate and for cleft palate (Woolf et al, 1964; Tanaka et al, 1967; Fujino et al, 1967; Carter, 1969).

It has been reported that a severely affected case would be more genetically predisposed than mild cases. Thus the frequency of the affected relatives would be more higher in the more severally affected cases (Carter, 1965).

Further studies may be made to test the specific hypotheses and making use of advantageous material such as syndromes involving cleft lip/palate, high risk families, and monozygous co-twins of patients. Observations on arch form, face shape, and body asymmetry's could be useful in helping to identify specific factors among those underlying the predisposition to clefts.

1.3 ASSOCIATED MALFORMATIONS, GENETIC DEFECTS AND ANOMALIES

Current literature suggests that there are well over 250 disorders associated with facial clefting (Gorlin et al, 1990). Several large clinic samples have been studied relative to the frequency with which clefts presents as one feature in pattern of malformations (Khoury, 1987; Rollnick, 1981; Sphrintzen, 1985). Rollnick and Pruzansky in 1981 retrospectively reviewed 4180 patients seen at the Center for Cranio Facial Anomalies at the University of Illinios. Multiple anomalies were identified in 35% of cleft lip and palate patients, 54% of cleft palate patients, and 55% of patients with submucous clefts. Review of 1000 patients evaluated at the Center for Cranio Facial Disorders at Montefiore Medical Center documented that 63% had an associated defect, with roughly half of these having a pattern of anomalies (Sphrintzen et al, 1985).

Knowing the different anomalies and malformations associated with cleft lip/palate enables the clinician to make proper diagnosis and this would provide a rapid way of sorting through the recognised syndromes with orofacial clefting a possibility for overall diagnosis. Through the diagnosis the frequency of clefting occurring with the syndrome, other features of the syndrome, and pertinent references can be known and understood.

A summary of few of the very commonly occurring anomalies associated with cleft lip/palate are provided in the Tables 1.3 and 1.4.

Table 1.3: Syndromes with cleft lip-palate

Syndrome	*Striking Features*	*References*
Appelt syndrome	Ocular hypertelorism, tetraphocomelia, enlarged penis or clitoris.	Appelt et al, 1966
Bixler syndrome	Hypertelorism, microtia, ectopic kidneys, congenital heart defect, growth deficiency	Bixler et al, 1969
Ectrodactyly-ectodermal dysplasia clefting syndrome	Ectrodactyly(hands and feet), sparse blond hair, oligodontia, nasolacrimal duct obstruction	Bixler et al, 1971
Hemifacial microsomia (Golden har syndrome)	Unilateral dysplastic ear, ear tags/pits, unilateral hypoplasia of mandibular ramus, and variably epibulbular dermoids, vertebral anomalies, cardiac defects, renal anomalies, other abnormalities.	Gorlin et al, 1976
Hypertelorism-hypo spadias syndrome	Hypertelorism, hypospadias, other abnormalities	Optiz et al, 1969
Juberg-Hayward syndrome	Microcephaly, hypoplastic distally placed thumbs, short radii	Juberg and Hayward, 1969
Meckel syndrome	Polydactaly, polycystic kidneys, encephalocele, cardiac anomalies other abnormalities	Hsia et al, 1971

Continuation of Table 1.3

Syndromes	*Striking Features*	*References*
Rapp-Hodgkin syndrome	Hypohidrosis, thin wiry hair, dystrophic nails	Rapp and Hodgkin, 1968
Van der Woude syndrome	Lip pits	Cervenka et al, 1967
Clefting/ectropion syndrome	Ocular hypertelorism, ectropion of lower eyelids, digital and/or limb reduction defects	Gorlin et al, 1971

Table-1.4: Syndromes with cleft palate

Syndromes	*Striking features*	*References*
Apert syndrome	Craniosynostosis, ocular hypertelorism, down slanting palpebral fissures, proptosis, midface deficiency, symmetric syndactyly of the hands and feet minimally involving digits 2,3, and 4, mental deficiency	Cohen, 1975
Christian syndrome	Craniosynostosis, microcephaly, arthrogryposis, adducted thumbs	Christian et al, 1971
Cleft palate/branchial plexus neuritis syndrome	Recurrent branchial plexus neuritis, limited extension at the elbows, facial asymmetry deep-set hypoteloric eyes.	Erickson, 1974
Cleidocranial dysplasia	Large calvaria, relatively small face, persistent fontanelles, supernumerary teeth, delayed eruption, absent or hypoplastic clavicles, other skeletal abnormalities	Gorlin et al, 1976
Ectrodactyly-cleft palate syndrome	Ectrodactyly and syndactyly	Optiz, 1975
Micrognathic dwarfism	Micromelic dwarfism, small mandible, cleft vertebrae	Maroteaux et al, 1970

Continuation of Table 1.4

Syndromes	*Striking Features*	*References*
Oro-facial-digital syndrome	Dystopia canthorum, hypoplastic alar cartilages, milia multiple frenula, laterally cleft palate, bifid tongue, malposed teeth, tooth anomalies, brachy dactyly, syndactyly, clinodactyly	Gorlin et al, 1976
Stickler syndrome	Myopia, retinal detachment, flat midface, prominent joints with degenerative joint disease	Hermann and Optiz, 1975
Wallace syndrome	Short limbs, deformed rib cage, hydrocephalus, hypoplastic lungs, congenital heart defects, central notch of upper lip	Wallace et al, 1970
Weaver-Williams syndrome	Mental deficiency, diminished subcutaneous tissue and muscle mass, microcephaly, hypoplastic ears midface hypoplasia, deep set eyes, small down turned mouth, malformed teeth, long thin neck, generalised bone hypoplasia, increased tubulation of long bones, delayed osseous maturation, down slopping ribs, clindodactaly.	Weaver and Williams, 1977
Foetal alcohol syndrome	Growth deficiency, mental deficiency, microcephaly, narrow palpebral fissures, congenital heart defects, joint anomalies, other abnormalities	Jones et al, 1973

Continuation of Table- 1.4

Syndromes	*Striking Features*	*References*
Kniest syndrome	Disproportionate dwarfism, round face, midface, short neck, lordosis, kyphoscoliosis, tibial bowing, progressively enlarged stiff and painful joints, clubfeet, sever myopia, retinal detachment, cataracts, deafness, recurrent respiratory infections	Siggers et al, 1974

It is important to have proper genetic evaluation in the treatment of children with clefts. The presence of "associated structural defects" in a child with a cleft is unlikely to be a random event, but rather should suggest the presence of a disorder with potential significance relative to prognosis and recurrence risk.

1.4 TREATMENT PROFILE FOR CLEFT LIP AND/OR PALATE AND CHANGES IN TREATMENT OVER TIME

The wide distribution of clefts in lower animals suggests that cleft of lip and/palate is a disease older than man himself. It is unfortunate that knowledge of early evolution of repair of the clefts is extremely sketchy. There is a vast amount of literature on cleft lip and/palate treatment today. The abundance of information in this field has created the impression that we know enough, or nearly enough, about the various treatment procedures developed by the individual disciplines to manage clefts successfully. But, an assessment the papers being published reveals that comparatively few of them are designed to evaluate the relative effectiveness of various treatment procedures through the use of proper methods of clinical research (Bardach, 1987).

(a). REPAIR OF CLEFT LIP

Undoubtedly the first sporadic attempts at approximating the raw edges of a cleft went unrecorded. Celsus, the first Century Roman dilettante who was an encyclopaedist and not a surgeon is credited with first description of a cleft lip repair (Garrison, 1968). According to Garrison (1968), the fourteenth Century Flemish surgeon Jean Yperman gave a good account of the healing of harelip, but Barsky (1964) considered Pierre Franco, a sixteenth Century French surgeon living in Switzerland, "to be the father of cleft lip surgery".

Despite the advances made by Renaissance surgeons, cleft lip repair remained crude, until the advent of anaesthesia and asepsis in the nineteenth Century permitted the development of complex incisions and refined suture techniques. Various surgeons contributed in advancement of surgical techniques among them the techniques introduced by Millard in 1955, Trauner and Trauner (1967), Wynn (1965), and Z-plasty's by Jayapathy, Huffman, and Lierly (1960). These are a few of the most accepted and widely followed techniques in repair of cleft lip today.

(b). TIMING OF CLEFT LIP REPAIR

Selection of suitable time for lip surgery varies from surgeon to surgeon and from clinic to clinic. In some parts of the world infants are operated upon under local anaesthesia during the first 48 hours of life. But "the rule of tens" is most widely advocated. This rule states that surgery should be delayed until the infants over 10 weeks of age, over 10 pounds in weight, and over 10 gms. in haemoglobin. Not only is the condition more suitable for surgery at such a stage, but also the lip and nose tissues have increased in size to facilitate the detailed surgery. Thus the age of three months has been most widely followed time for the initial lip surgery (Millard, 1979; Wilhelmsen and Musgrave, 1966).

(c). REPAIR OF CLEFT PALATE

The treatment of cleft palate has undergone evolutionary changes. From the use of a prosthetic obturator to the surgical closure of the palatal clefts, many individuals have contributed to the advancement of cleft palate management. Graefe and Roux (1819) have been credited with the earliest successful closure of soft palate cleft. Veau (1931), Kilner (1937), Wardill (1937) also made remarkable contribution in improving the techniques (Peet, 1961).

Schweckendiek in 1978 advocated the use of a two stage cleft palate closure. The soft palate was closed early, with closure of the hard palate delayed until several years later. The rationale for the two stage closure was to provide improved velopharengeal function during initial speech development by the early closure of the soft palate. The hard palate closure was delayed to allow the hard palatal cleft to narrow with facial growth, to facilitate surgical closure and most of all to minimise the deleterious effect of surgery on facial growth (Schweckendick, 1978).

Several techniques for primary veloplasty have been proposed. The historical development of the management strategy proceeded along the following theme: obturation of the cleft, total closure of cleft palate, lengthening of the palate, two stage closure of cleft palate, the anatomic velopharengeal muscle

repair. These historical developments underlie the existing controversies in the cleft palate management.

(d). TIMING OF CLEFT PALATE REPAIR

Surgical interventions in cleft palate improved some aspects of cleft palate pathology, but can cause a host of unwanted sequelae. The management strategy of cleft palate has several areas of controversy:

(1) timing of surgical intervention;

(2) types of surgical repair; and

(3) orthodontic management.

These appear to be separate concerns, but in reality they are interrelated. A comprehensive understanding is necessary to develop a coherent treatment plan.

Early in the development of surgical techniques in cleft palate repair, the main interest was to obtain closure. Various kinds of ingenious methods of tissue manipulation were developed to achieve closure without the recognition of the possible facial growth problems inherent in the surgical interventions. As these patients grow, the facial, dental and speech deformities become apparent. To avoid these problems other techniques were developed.

Schweckendiek advocated the two stage cleft palate closure. He proposed that closure of the soft palate be performed at 6-8 months, with hard palate closure delayed until 12-14 years of age (Schweckendiek, 1978).

Dorf and Curtin in their patients advocated a primary single stage repair after 12 months of age (Dorf and Curtin, 1982). Other authors have proposed different timing for the two stage closure. Rohrich and Byrd recommended closure of the soft palate at 3-6 months, followed by hard palate closure at 15-18 months. The rationale was to facilitate the closure of the hard palate, because the palatal cleft would narrow by 15-18 months(Rohrich and Byrd, 1980). Witzel et al (1984) in their review of the available data on delayed hard palate closure, however, found no convincing evidence that the delayed hard palate closure, if done prior to full facial growth, would reduce that amount of facial and dental distortion. There is an isolated report of acceptable speech development with delayed hard palatal closure(Perko, 1990). The list of surgical techniques used in palatal cleft closure is extensive. Due to unavailability of any long term studies analysing single treatment procedure it is very difficult to comment on any one single procedure or surgical intervention.

(e). PRIMARY ALVEOLAR BONE GRAFTING

Primary or early bone grafting consists of placing a bone graft at the cleft site before the eruption of deciduous teeth or before the age of one year has been

reached. A few reports on early bone grafting appeared at the beginning of this Century, but by the 1950s and 1960s a lot of reports on bone grafts in the area of alveolus were published.

(f). TIMING FOR BONE GRAFTING

A review of earlier reports revealed wide variation in the timing of graft placement, the source of graft, the location of graft placement and the extent of dissection performed. The time ranges from the age of 4-9 months of age (Brauer et al, 1962; Schuchartt et al, 1966).

(g). SECONDARY ALVEOLAR BONE GRAFTING

Secondary bone grafting of the maxilla and the residual alveolar clefts in conjunction with orthodontic treatment are generally carried out at the stage of the transition dentition. It has been incorporated in the management protocol of many cleft / cranio facial teams as an adjunct procedure aiming to further improve the functional and aesthetic outcome of patients with unilateral or bilateral cleft of lip and palate (Bergland et al, 1986; Semb, 1991).Bone grafting of residual alveolar clefts was first reported by Von Eiselsberg in 1901, followed by Lexer in 1908 and Drachter in 1914.

These procedures got their acceptance only in 1950s. With accumulated experience and long term evaluation and follow up, several benefits of the procedure were recognised (Robertson et al, 1983; Abyholm et al, 1981).

(h). TIMING FOR BONE GRAFTING

Currently there are two schools of thought regarding the timing of bone grafting:

(1). early bone grafting during or shortly after and full orthopaedic alignment of the maxillary segments (Nylen et al, 1974; Rosenstein et al, 1982);

(2). secondary bone grafting at the stage of the transitional dentition prior to the eruption of the permanent canine or even the lateral incisor, in conjunction with orthodontic treatment (Enemark et al, 1987; Eskeland et al, 1985).

Internationally, the majority of surgeons currently favour bone grafting of residual alveolar clefts at the stage of the transitional dentition and agree that, with close cooperation between the surgeon and the orthodontist, good repeatable results can be obtained.

(I). OTHER TREATMENT VARIABLES

Treatment of cleft lip and/or palate involves other surgical and medical variables such as speech and hearing development, orthognathic surgery,

orthodontic treatment, correction of secondary lip and nose deformity, correction of velopharengeal insufficiency and secondary palatal management, etc. All these interventions are carried out at various times, depending on the surgeon and the type of defect.

(j). SPEECH AND VELOPHARENGEAL INSUFFICIENCY

Normally speech therapy starts by the time the child is 12 months of age and then regular assessment at various intervals followed by intervention appropriate to the age is implemented. Normal children usually speak their first word by the age of 13 months. However, the initial babbling that usually begins prior to this stage is part of continuum of speech development. There appears to be a continuity of speech development from prelinguistic babbling at 6-9 months of age to later stages of speech development. The range and the evolution of the babbling sounds have implications in the future speech development (O'gara et al, 1990; Zimmerman and Canfield, 1968). Competent velopharengeal function is believed to be very important during this period. If there is significant velopharyngeal insufficiency, the abnormal and often irreversible secondary compensatory speech, such as glottal and pharyngeal articulation may develop. At this stage a Pharyngoplasty is carried out to provide velopharyngeal competency in patients (Skolnick, 1969; Hogan, 1973).

(k). ORTHODONTIC - SURGICAL INTERVENTION

The timing and sequencing of treatment require close collaboration of the team. Deciding to delay surgical orthodontic treatment until growth is stabilised may be sound judgement, but not always in patient's best interest, especially when psychosocial development is affected. As a general rule, skeletal surgery, orthodontic intervention, and final prosthetic rehabilitation should be completed before soft tissue revisions or rhinoplasty are instituted. In general orthodontic correction is brought about during the estimated time for growth spurts and the surgery follows after the completion of the growth spurt (Subtelny, 1990; Bardach et al, 1990; Proffit, 1993).

(l). CORRECTION OF RESIDUAL DEFORMITIES OF THE LIP AND NOSE

The secondary reconstruction cleft deformities begins by providing a bony foundation for the lip and nose. Secondary bone grafts, orthognathic surgery and orthodontic treatment should all be completed. Because of the swelling and distortion of soft tissues accompanying the procedures for construction of bony foundation, the definitive lip and nasal reconstruction is usually carried out separately, 3-6 months later (Jackson et al, 1990; Mulliken, 1992).

1.5 TREATMENT PROFILES AT OTHER CENTERS

Inter-center studies offer tremendous advantages in cleft lip and / palate research as they allow direct comparison of the outcome of primary surgery together with other major components of the treatment program in respective centers. Very few investigations on single center or multi-center treatment outcomes have been done.

A recent review of *The Cleft Palate Journal* revealed only six such studies in previous 24 years have been done (Shaw et al, 1992). A single center study was spotted in Clinics in Plastic Surgery (Witt et al, 1993). There are about 117 identified reports on some aspect of treatment outcome, most of which were on facial growth and dental occlusion and one on speech pathology (Shaw et al, 1993).

After the Fifth International Congress on Cleft Lip and Palate and Related Cranio facial Anomalies in 1986, the European Cleft Lip and Palate Research Group was formed by orthodontists of six participating centers. This research group developed a series of aims, but its main focus was a comparative study of treatment outcome in unilateral cleft lip and palate patients from the six centers (Molsted et al, 1993). The initial results of these participating groups have now been published (Asher- McDade et al, 1992; Mars et al, 1992; Molsted et al, 1992; Shaw et al, 1992 a, b).

The treatment followed by the participating centers is described in Table 1.5

TABLE 1.5: Treatment protocol of the six participating centers (Shaw et al; 1992)

AGE	A	B	C	D	E	F
Birth	Presurgical orthopaedics (HOTZ)		Presurgical	orthopaedics (Extra oral strapping)	Presurgical	orthopaedics (T-traction)
3 mo	Lip closure (Millard, Skoog)	Lip closure (Tennison & Vomerplasty)	Lip closure (Variation of methods & timing)	Lip closure (Variation of methods & timing)	Lip closure (Millard & Vomerplasty)	Lip closure (Modified Skoog, Tennison & Al bone graft
6 mo						
9 mo	Soft palate closure(Von Langenbeck, Perko, Wardill Kriens)					
12 mo			Palate closure (Variation of method & timing)	Palate closure (Variation of methods & timing)		Palate closure (Veau-Wardill Kilner)
18 mo					Palate closure (Modified Von Langenbeck)	
24 mo		Palate closure (Wardill pushback)				
3 yrs						
6 yrs						
9 yrs	Bone grafting (Hard palate closure)	Bone grafting	Bone grafting	bone grafting	Bone grafting (only if failure)	Bone graft

SUMMARY OF TREATMENT AT EACH OF THE CENTERS

Each of the six centers will be identified by a letter, A - F. The main features of the treatment in each center are indicated in the previous table and described below.

CENTER A:

The surgical management at this center consisted of lip closure at 3 - 4 months of age. Most commonly a Millard procedure was used, and few cases had a Skoog procedure. At the age of 9 -15 months, the soft palate closure was carried out, using the Von Langenbeck, Perko, Wardill or Kriens procedure. The closure of hard palate normally was done at the age of nine years and in some cases at the age of six years. Presurgical orthopaedics was used by this center.

CENTER B:

The surgical management consisted of lip closure at two months of age and they used Tennison procedure, and at the same time vomerplasty was done to close the anterior part of the hard palate. The rest of the palate was closed at 22 months of age by a Wardill push back procedure. All operations were performed by the same surgeon.

CENTER C:

The surgical management varied as many different surgeons were involved in the primary procedures. But in general the lip was closed within the first six months of age and palate was closed at approximately one year of age.

CENTER D:

In this center many different surgeons were involved in the primary surgery. But in general the lip was closed within the first six moths of age and the palate was closed within the first two years of age. Presurgical orthopaedics with extraoral strapping was used.

CENTER E :

At this center the lip closure took place at the age of three months using a Millard procedure. At the same time anterior palate was closed by vomer-plasty. The rest of the palate was closed at the age of 18 - 20 months using modified Langenbeck procedure.

CENTER F:

This center used presurgical orthopaedics with extraoral traction for correction of the septal deviation and primary alveolar bone grafting with macerated rib. Lip closure was done at 4 -6 months of age, using mostly a

modified Skoog procedure or a Tennison/Randall procedure. The palate was closed at 12 months of age using a Veau-Wardill Kilner procedure.

No orthodontic treatment was carried out in the deciduous dentition in any of the centers. In mixed dentition simple correction of malocclusion prior to secondary bone grafting was used in all the centers. Center F used secondary bone grafting in the cases where the primary bone grafting had failed. All other centers used secondary bone grafting.

Another isolated study found was by the Loma Linda University Cranifacial team, describing the protocol which was adopted during the last six years (Witt and Robert, 1993). The principal features of treatment in this center are indicated in the Table 1.6.

TABLE - 1.6
Loma Linda University Craniofacial team protocol for cleft lip and palate

Age	Clefting of primary palate only: Incomplete Cleft Lip	Clefting of primary palate only: Complete Lip Primary Palate	Clefting of Primary & Secondary Palate	Clefting of Secondary Palate
Birth	Complete cranio facial team evaluation	Complete cranio facial team evaluation	Complete cranio facial team evaluation	Complete cranio facial team evaluation
6 weeks		Cleft lip adhesion	Cleft lip adhesion Dental impression Insertion of passive molding plate	
3 mo	Definitive cleft lip repair			
6 mo		Definitive cleft lip repair	Interim dental impressions Definitive cleft lip repair	
1 yr	Complete cranio facial team evaluation	Complete cranio facial team evaluation	Complete cranio facial team evaluation Interim dental impressions Palatoplasty	Complete crano facial team evaluation Palatoplasty
2 yrs	Yearly complete team evaluation	Yearly complete cranio facial team	Yearly complete cranio facial team evaluation	Yearly complete cranio facial team evaluation
3 yrs			Yearly evaluation and secondary management of velopharengeal dysfunction (as needed)	Yearly evaluation & secondary management of velopharengeal dysfunction (as needed)
5-7 yrs		Alveolar bone grafting	Alveolar bone grafting	
13-18 yrs		Rhinoplasty (as needed)	Rhinoplasty(as needed) Orthognathic surgery (as needed)	

(Witt and Robert 1993)

There has been little in the way of inter-center collaboration, except for the meeting and attendances at the formal conferences. Thus even for durable records such as cephalograms and study casts, highly varied systems of analysis have been employed. This generally precludes detailed and statistically reliable comparison of one published report with another (Semb, 1991).

Retrospective study requires information on primary management practices 5,10 or 20 years previously and presupposes clearly described treatment protocols (Shaw et al, 1992).

1.6 RATIONALE FOR THE PRESENT STUDY

There is an abundance of literature on cleft lip and palate treatment available today. This creates an impression enough is known about the various treatment procedures developed by the individual disciplines to manage clefts successfully. Their is also the impression that the various treatment procedures have been carefully investigated and that teams are now able to consistently get satisfactory results. Lastly it is assumed that multidisciplinary management, interaction between various specialists, and the functioning of the cleft palate team are all well established and analysed from different aspects. But, very few published papers are designed to evaluate the relative effectiveness of various treatment procedures through the use of proper methods of clinical research. There is no universally excepted treatment procedure or plan for the various clefts (Bardach et al, 1984).

It is very difficult to find any well documented, long term results analysing single treatment procedure. It is amazing that, with so many well established cleft palate teams and cleft palate centers, there are very few studies conducted to analyse the effectiveness of multidisciplinary treatment. Studies within a single institution as well as comparative studies

between two or more institutions regarding the patient management and treatment procedure and profile are strangely scarce.

There may be several reasons to why there are so few analyses of treatment profiles or multidisciplinary treatment strategies. One answer to this to this question may be people are embarrassed at the prospect of reporting results which fall short of expectations, while these expectations are, perhaps, kept artificially high by the absence of reports. But in the interest of serving patients as well as possible, of understanding what is done and of improving techniques, timing and sequelae as much as possible, it seems it is necessary to begin a discussion of the problems encountered as well as the triumphs achieved. This is the rationale of the present study. Its purpose was to evaluate the mutidisciplinary approach towards treatment of facial clefts by the Australian Cranio Facial unit at the Adelaide Children's Hospital and the change in treatment profile over time.
By investigating and analysing the advantages and disadvantages of treatment profiles followed at various Cranio Facial units, it would be possible to establish more reliable criteria for the state of art of the field. The critical evaluation of the results achieved by various teams using their treatment profile must be based on properly designed, scientific foundations to make the conclusions warranted. And this may stimulate the

development of improved treatment strategies and techniques which would benefit cleft lip and palate patients (Bardach, 1987).

1.7 STUDY OBJECTIVES

Cleft lip and palate is one of the most common congenital anomalies. It presents a serious problem to the health delivery system because prevention seems very unlikely in the predictable future, as the aetiology is not yet known precisely. At present our knowledge of the tetragons that are associated with clefting is very limited. Genetic counselling can identify high risk patients, and ultrasound at 18 weeks gestation can reveal markers of syndromes associated with clefts. The clefts themselves may be visible at 20 weeks. But beyond early identification, we can only look to the future (McComb, 1989; Ross, 1987).

Multidisciplinary treatment, will be necessary for many years to come. Therefore, it is imperative that efforts be redoubled to improve treatment strategies and techniques to obtain better overall results. For this long term studies are requiered analyse treatment strategies and techniques at various Cranio Facial Units.

The main objective of this study was to provide information and data regarding the multidisciplinary approach and treatment strategies followed at the Australian Cranio Facial Unit in correction of various facial clefts

and change in treatment strategies over a period of time. This will enable the multidisciplinary team to evaluate their treatment profile for various types of clefts. This will enable the unit to adopt a sequence of repair so as to get maximum results and to reduce patient hospitalisation and medical care cost.

This study also aimed in provide well documented data so as to help future researches to evaluate the treatment stratergies at the Australian Cranio Facial Unit on a single center basis or for multicenter studies. The data also provide some measures against which other centers might evaluate their own treatment profiles. This study will enable differences to be found in treatment profiles followed at other centers. These differences may be small or large, but may have a great impact on the future habitation of the children. Such a finding would be of importance in formulation of treatment strategies beneficial to the multidisciplinary team, and patient and society.

The last objective of this study was to provide data from which hypotheses for more rigorous research and testing could be generated which would bring about more comprehensive, coordinated care provided by the health care system that would be readily accessible and responsive to individual needs of the patients and their families. Among these other points are :

1. facilitation of parent/professional collaboration in the health care of children;
2. sharing of unbiased and complete information about children with their parents;
3. provision of emotional and financial support to families;
4. sensitivity to cultural differences;
5. encouragement of parent to parent support;
6. incorporation of the developmental needs of infants, children and adolescents into health care plans;
7. assurance of the availability of comprehensive services including social, emotional, and cognitive aspects of health care; and
8. an interdisciplinary approach to care.

(Tullock 1993; American Cleft Palate-Cranio Facial Association, 1993; Surgeon General, 1987).

2 MATERIALS AND METHODS

2.1 STUDY POPULATION

A excellent opportunity exists in South Australia for study or research in the field of cleft lip and palate. With a very few exceptions, most of the cleft lip and / or palate cases are referred to the Adelaide Children's Hospital for treatment. The reason for this is that feeding difficulties and other postnatal problems in the care of cleft affected children require specialist nursing training and facilities. In addition , the child affected with cleft requires a multidisciplinary approach for compete treatment of the cleft and other associated defects. The Adelaide Children's Hospital has a very well established Australian Cranio Facial Unit which comprises of staff of interdisciplinary team and all the team members are trained and experienced in the care of patients with cranio facial anomalies.

The Medical Records Department of the Adelaide Children's Hospital maintains an index, based on the medical condition responsible for all admissions. Admissions with multiple anomalies and with an oral or facial cleft as a secondary condition were also listed. In addition, the Australian Cranio Facial Unit also maintains all the records of patients who have been referred for treatment of facial clefts.

The pertinent records were collected and the name, hospital record number and cranio facial unit number were noted down of all the children admitted for surgery over the years 1940 to 1993.

The hospital case notes of the listed admissions were recorded on a year to year basis, and the subjects were included for the study when the following conditions were fulfilled :-

1. case notes of the particular patient were located both at the Medical Records Department of the Hospital as well at the Cranio facial Unit of the hospital;
2. the subject was born between 1940 to 1993;
3. the subject had cleft of lip and /or palate as a whole entity or associated with other defects;
4. all the subject selected must have completed all the primary surgical interventions; and
5. the subject had completed all treatment (surgical interventions for the cleft) at the Adelaide Children's Hospital.

All subjects included for further study were listed alphabetically by surname, and a study number was given. Perusal of hospital case notes yielded data of a varying degree of completeness for individual subjects. In spite of this limitation, basic study data were obtained in combination from the case notes from the Medical Records Department and Cranio Facial

Unit on all subjects and recorded on a field record form CFU I (Appendix A1). Complete case notes were necessary so as abstract the exact dates and times of hospital admission for various surgeries.

The Adelaide Children's Hospital lists over 750 children with clefts of the orofacial complex. As 255 of these fulfilled the study requirements for this report, they were given a study number.

The following subjects were excluded from the study

1. deceased after a few months of birth;
2. case notes not found in either Medical Records or Cranio Facial Unit;
3. incomplete case notes;
4. admitted for elective surgery (eg: patient from overseas who was admitted for a particular surgery);
5. other type of facial clefts (other than cleft lip and /or palate);
6. subjects born before 1940 and after 1993; and
7. incomplete treatment for whatever reason.

A protocol was submitted to the Adelaide's Children's Hospital Research Ethics Committee for permission of the study and at the same time the Medical Records Department and Cranio Facial Unit. Permission to assess the patients case notes for the study was requested from them. The

permission was granted and the case notes of subjects were studied thoroughly and the required details were abstracted.

2.2 DATA COLLECTION AND PREPARATION

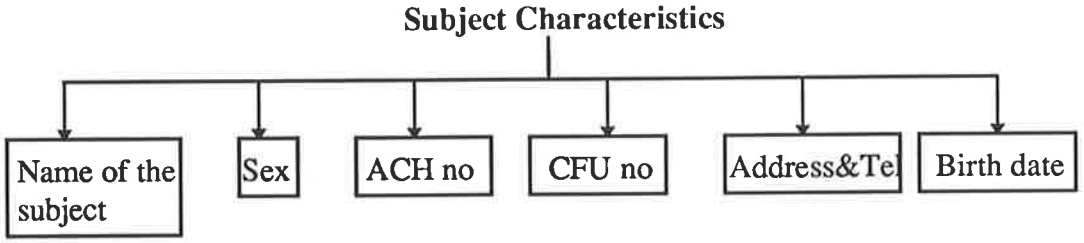
In consultation with Ms Joan Alister of the Department of Dentistry, The University of Adelaide, a data collection sheet was prepared CFU I (Appendix A1), to enter the details required from the subjects case notes for the study.

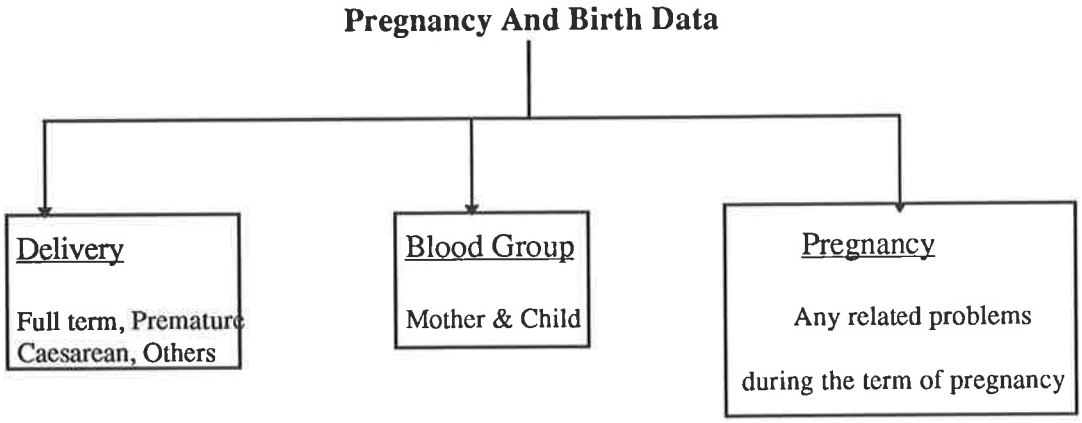
Medical Records Department of Adelaide's Children's Hospital was very cooperative during the time of the study. The administrator of the Medical Records Department provided a small area where case notes could be studied and the staff helped in removal of the case notes from various storage sections of the Hospital.

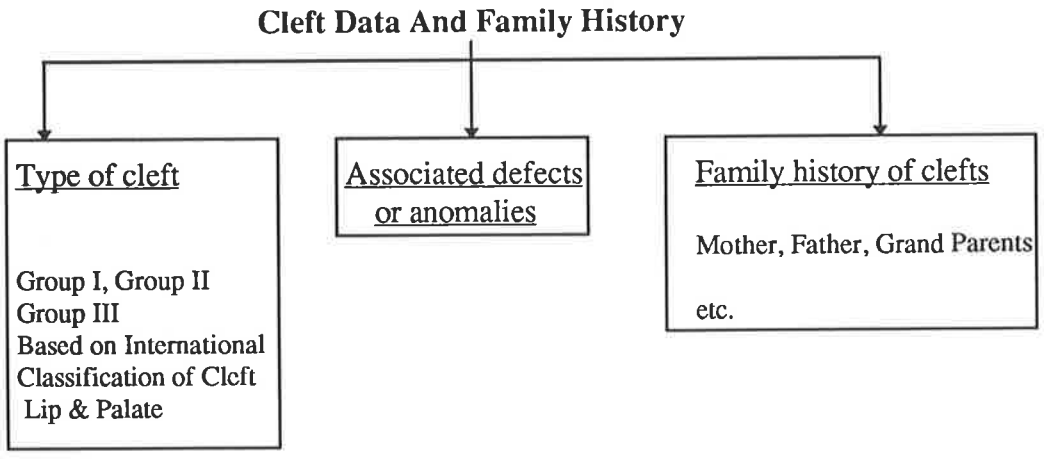
2.3 DATA ITEMS

Once the case notes of the particular subject were taken out the required information was noted down on the prepared CFU I form prepared.

The following items were abstracted from the case notes of selected study subjects.


Subject Characteristics
Name of the subject
Sex
ACH no
CFU no
Address&Tel
Birth date


Pregnancy And Birth Data
Delivery
Full term, Premature Caesarean, Others
Blood Group
Mother & Child
Pregnancy
Any related problems
during the term of pregnancy


Cleft Data And Family History
Type of cleft
Group I, Group II
Group III
Based on International
Classification of Cleft
Lip & Palate
Associated defects or anomalies
Family history of clefts
Mother, Father, Grand Parents
etc.

Surgical Interventions & Timing

- Lip Repair → Age of 1st repair
  → No. of secondary lip repair
- Palate Repair → Age of 1st repair
  → No. of secondary palate repairs
- Alveolar Bone Grafting → Age of 1st graft
  → No. of secondary bone grafts
- Pharyngoplasty → Age of 1st repair
  → No. of secondary repairs
- Rhinoplasty → Age of 1st repair
  → No. of secondary repairs
- Bilateral Myringotomies → Age of 1st insertion
  → No. of secondary insertion
- Osteotomies → Age at the time of intervention

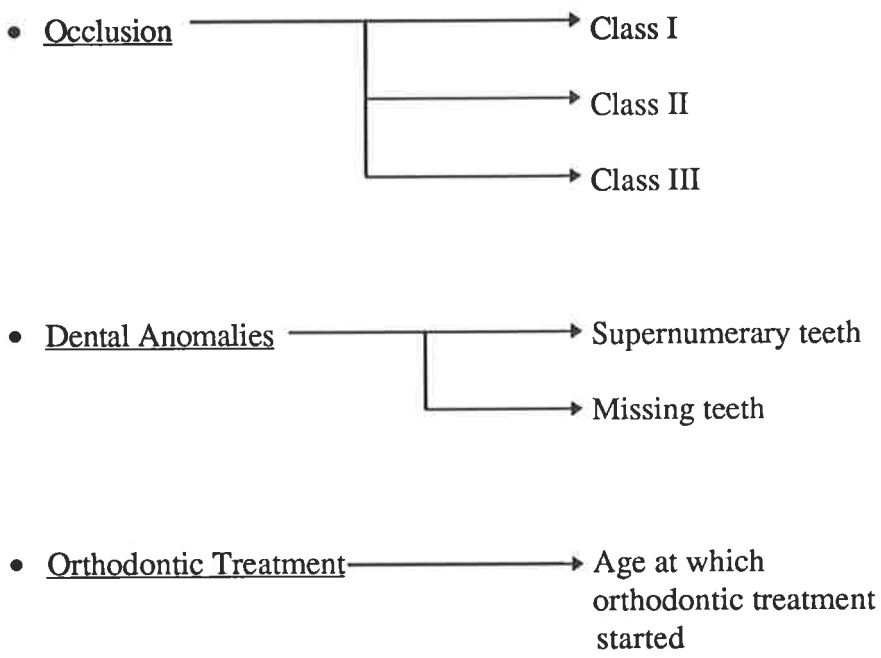
The notes and dates of the surgical intervention were taken down in proper sequence. If there were any other surgical interventions related to the correction of a cleft, these were also taken down and the date at which the surgery was performed noted. The total number of days of hospitalisation for the surgeries were calculated by noting down the day of hospitalisation and day of discharge.

MEDICAL INTERVENTIONS

- Speech therapy → Age at which started
  → Approximate number of therapy sessions
- Nasendoscopy → Done or Not
- E . N. T Treatment → Done or not
- Radiographs and photography → Approximate number of occasions
- Genetic counselling → Done or Not

Apart from the above reasons other medical conditions were noted down if the subject was admitted to the Hospital for treatment eg. asthma, chest infection, dermatitis etc.

Dental Interventions And Defects



It has been recognised that any retrospective review of hospital records and patient examination many years following a repair presents difficulties in ascertaining the original anatomical extent of the defect. For this reason, analysis of completeness or severity of the cleft condition was not attempted.

2.4 DATA ANALYSIS

The data were transferred from the form CFU I (Appendix A1) to a computer data file at Department of Dentistry, the University Of Adelaide. In consultation with Ms. Joan Alister of the Department of Dentistry, The University of Adelaide, programs were written to enter the data collected in the computer and the data was analysed using SPSS package. Where appropriate the results are presented in graphical form using Harward Graphics.

3: RESULTS

3.1 SAMPLE CHARACTERISTICS

There are 750 cases registered at the Adelaide Children's Hospital for treatment of clefts of orofacial complex of which only 255 cases were incorporated into the present study. Case selection was done on basis of certain criteria. The following cases were excluded from the study:

1. deceased after a few months of birth;
2. case notes not found or missing;
3. incomplete case notes;
4. admitted for elective surgery;
5. other types of facial clefts; and
6. incomplete treatment.

Out of 792 cases, 190 cases were excluded from the present study as these cases were admitted for treatment of various other facial clefts, eg, tessier clefts. Twelve(12) cases were deceased after a few months of birth. Two hundred and twelve (212) cases were admitted for elective surgery, most of these cases were from overseas or interstate hence were not incorporated in the study. Thirty-eight (38) cases were excluded from the study as they did not complete their treatment, due to migration to different State or

country. Case notes of 115 subjects were incomplete, having missing or improper data recorded and hence excluded from the present study. Hence, the results of the present study were based on 255 subjects who satisfied the study criteria.

TYPE OF CLEFTS

Among the 255 case selected for the study, clefts of anterior (primary) palate made up to 27.5% of the total (Group I), clefts of posterior (secondary) palate 23.9% (Group III), and combined clefts of the anterior (primary) and posterior (secondary) palate contributed 48.6% (Group II). The distribution is shown in the Figure 3.1 below. Cleft classification was done on basis of International Classification of Cleft Lip, Alveolus and Palate (Hall, 1994).

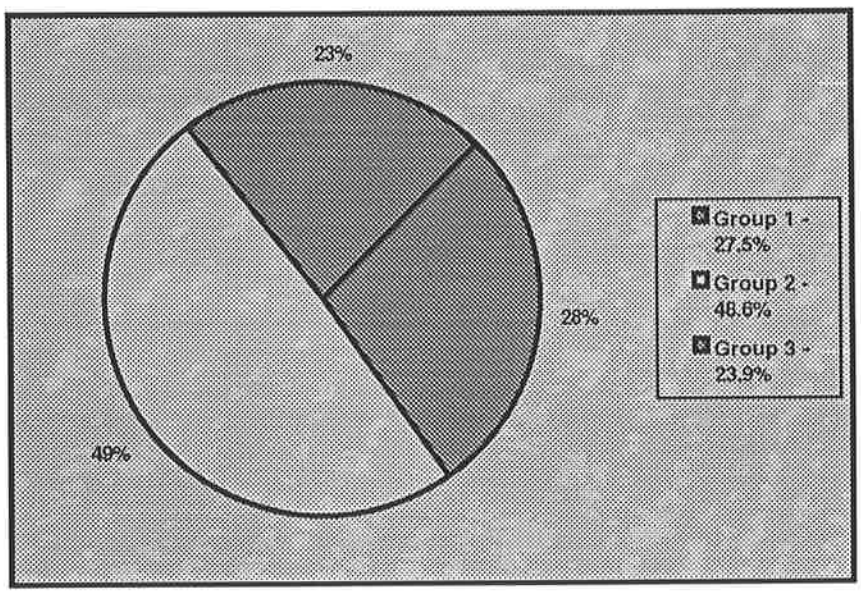


FIGURE 3.1 Type of cleft and population

SEX RATIO

There were more males than females. Out of the study population of 255 subjects 94 were females and 161 were males .

Female : male ratio was found to be 1 : 1.71. The percentage distribution is represented in Figure 3.2.

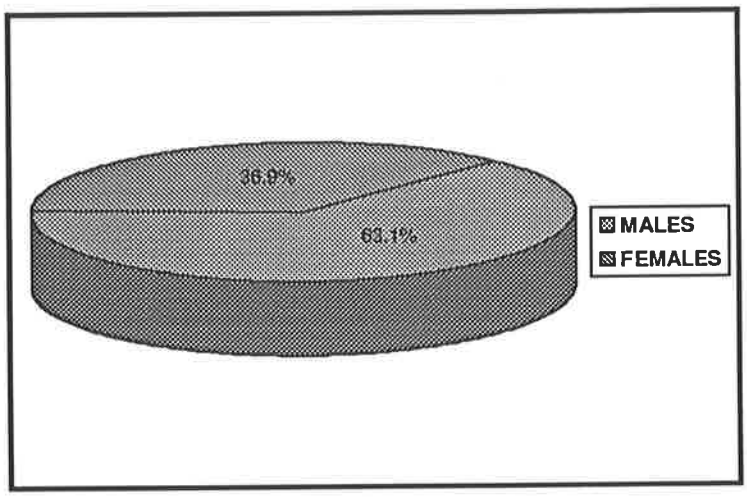


FIGURE 3.2 : Sex distribution for cleft lip and palate

FAMILY HISTORY

No information on family history of clefts was available for 218 of the 255 subjects. Table 1.7 gives the number of probands in subjects with known family history and subjects with no family history, and percentage of

probands with known affected "near relatives" and also "all known" affected relatives. The "near relative" group included the first five nearest sets of relatives (siblings, parents, aunts and uncles, grandparents and first cousins). The "all known" relative group included near relatives and others of more distant yet known relationship to the cleft affected proband.

TABLE 1.7: The proportions of proband with family history of cleft among near and all known relatives.

Subjects with no family history information	Subjects with positive family History	Family history positive near relatives		Family history positive all known relatives	
		No.	%	No.	%
218 (85. 5%)	37 (14. 5%)	23	9%	14	5. 5%

FAMILY HISTORY OF OTHER GENETIC DEFECTS

Out of 255 subjects 36 subjects reported to have family history of other genetic defects, eg, congenital heart disease, mental retardation, diabetes. Some cases reported no information on family history of any other genetic defect or anomalies.

BLOOD GROUPING

Blood group details of 82 cases were missing. The blood grouping abstracted form the remaining cases are represented in the Figure 3.3 below.

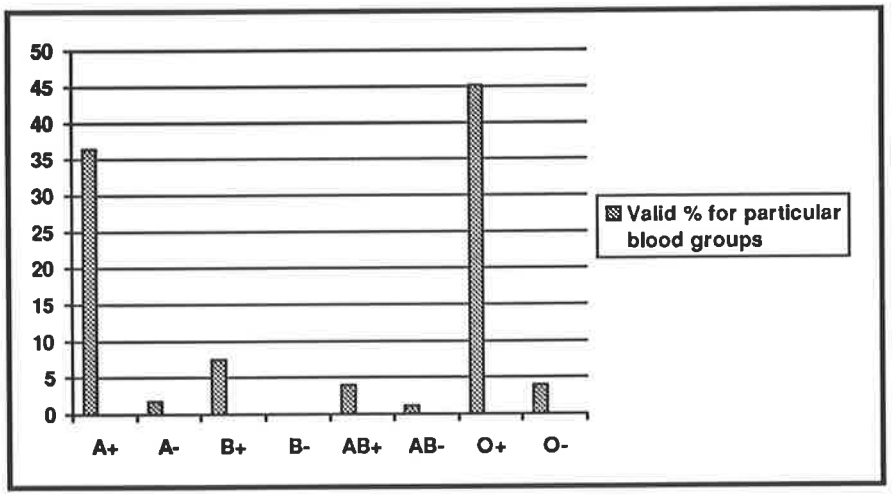


FIGURE 3.3 : Distribution of blood grouping

The study sample had a high percentage of A+ blood type (36.4%) and O+ blood type (45.1%) (Appendix B).

3.2 TREATMENT DATA

The surgical procedures which are commonly employed in treatment of cleft lip and / or palate included in the present study are as follows

1. repair of cleft lip;
2. repair of cleft palate;
3. alveolar bone grafting;
4. pharyngoplasty;
5. rhinoplasty;
6. bilateral myringotomies; and
7. osteotomies.

The medical intervention included in the study is speech therapy and the dental treatment included in the study is orthodontic intervention.

Emphasis is given to these few intervention, as these are the main interventions which bring about the changes in facial appearances, muscular balance, oro-facial growth, speech development and improvement, and normal occlusion development. Any any small changes such as timing or procedure in conducting these interventions could go a long way in bringing about desirable results. Most of the interventions employed in treatment of cleft lip and / or palate as whole entity are interdependent, and

change or failure in one intervention could directly or indirectly effect the other following interventions.

The study group was divided into 5 groups according to cohort defined as those born in (a) 1940 -1959 ; (b) 1960 -1969;

(c) 1970 -1979; (d) 1980 -1989; and

(e) 1990 -1993.

and dates of first interventions for all the listed surgical, medical and dental procedures were extracted, and analysis was done using the SPSS computer package and the mean age and standard deviation of each procedure in a particular cohort group was derived. These are represented in graphical form, where each unit of X axis represents a decade and each unit of Y axis represents age in years.

3.2.1 MEAN AGE OF FIRST INTERVENTION OF VARIOUS INTERVENTIONS BY COHORT AND CHANGES OVER TIME

REPAIR OF CLEFT LIP

Figure 3.4 shows that mean age of intervention for repair of cleft lip has decreased from the 1960s cohort to the 1970s cohort. In the last three birth cohorts, the mean age of intervention has been similar, ie, age at the intervention has been between 3 - 6 months. But the deviation around the mean age has significantly decreased. In the 1990s birth cohort there was a relatively very small deviation aroun the mean which implies that the all interventions were done at the age of 3 - 4 months (Appendix C1).

REPAIR OF CLEFT PALATE

As seen in Figure 3.5, there was a trend of decrease in mean age at intervention across all birth cohorts. The deviation from the mean age has also decreased. This implies that for the 1990s birth cohort most of the interventions were carried out in between 6-12 months of age (Appendix C2).

Figure 3.4: Mean age of first intervention - Repair of cleft lip

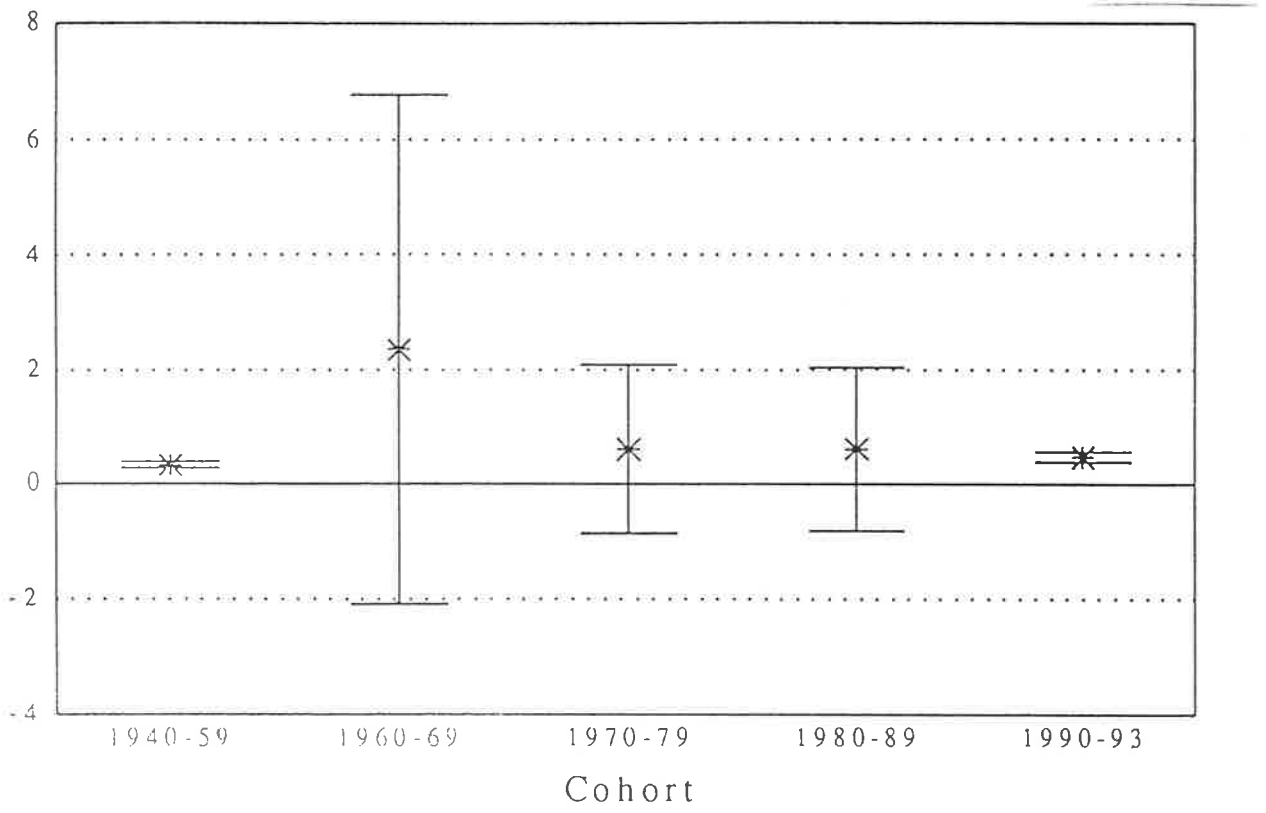
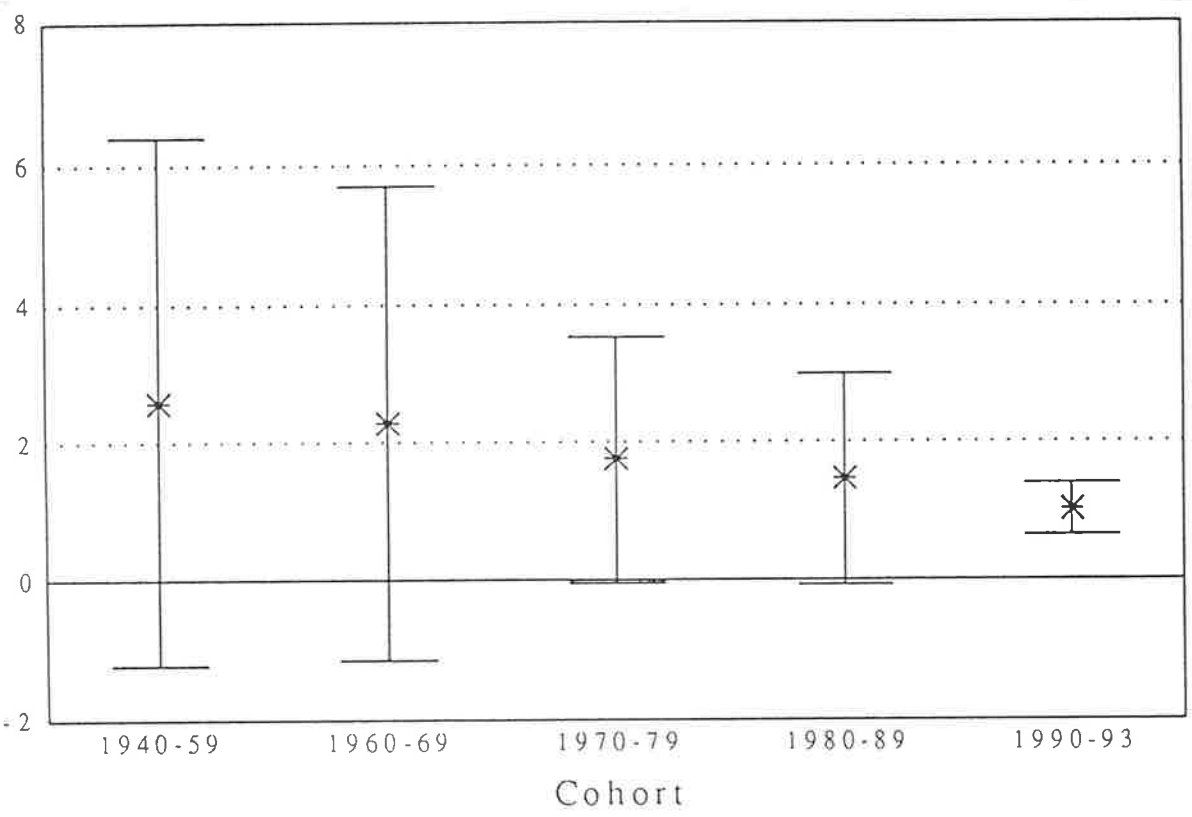


Figure 3.5: Mean age of first intervention - Repair of cleft palate

Age of intervention (Anova ; $p < 0.05$)



ALVEOLAR BONE GRAFT

As seen in Figure 3.6, the mean age at intervention has very substantially decreased since the 1940s birth cohort. In particular, the mean age showed a dramatic decrease for the 1960s cohort. Since then there has been a constant, but small decrease until the 1990s. The deviation around the mean has also constantly decreased across the last three cohorts. Most of these interventions by the year 1980 and onwards were done at 10 - 12 years of age (Appendix C3).

PHARYNGOPLASTY

As seen in Figure 3.7, the mean age of intervention has significantly decreased in the last five decades. The deviation from the mean has also been considerably decreased. By the year 1980 and onwards, most of the interventions were done between the age of four to nine years. (Appendix C4)

RHINOPLASTY\ NASAL TIP REVISION

From Figure 3.8, it can be observed that the mean age of intervention has decreased across the five birth cohorts. The mean age of intervention has been relatively constant since the 1970s cohort onwards. The age of intervention is 9 -12 years. The standard deviation initially decreased, but it increased for the 1970s cohort and later it decreased again to quite low valuefor the 1980s cohort.

Figure 3.6: Mean age of intervention - Alveolar bone graft

Age of intervention (Anova ; $p > 0.05$)

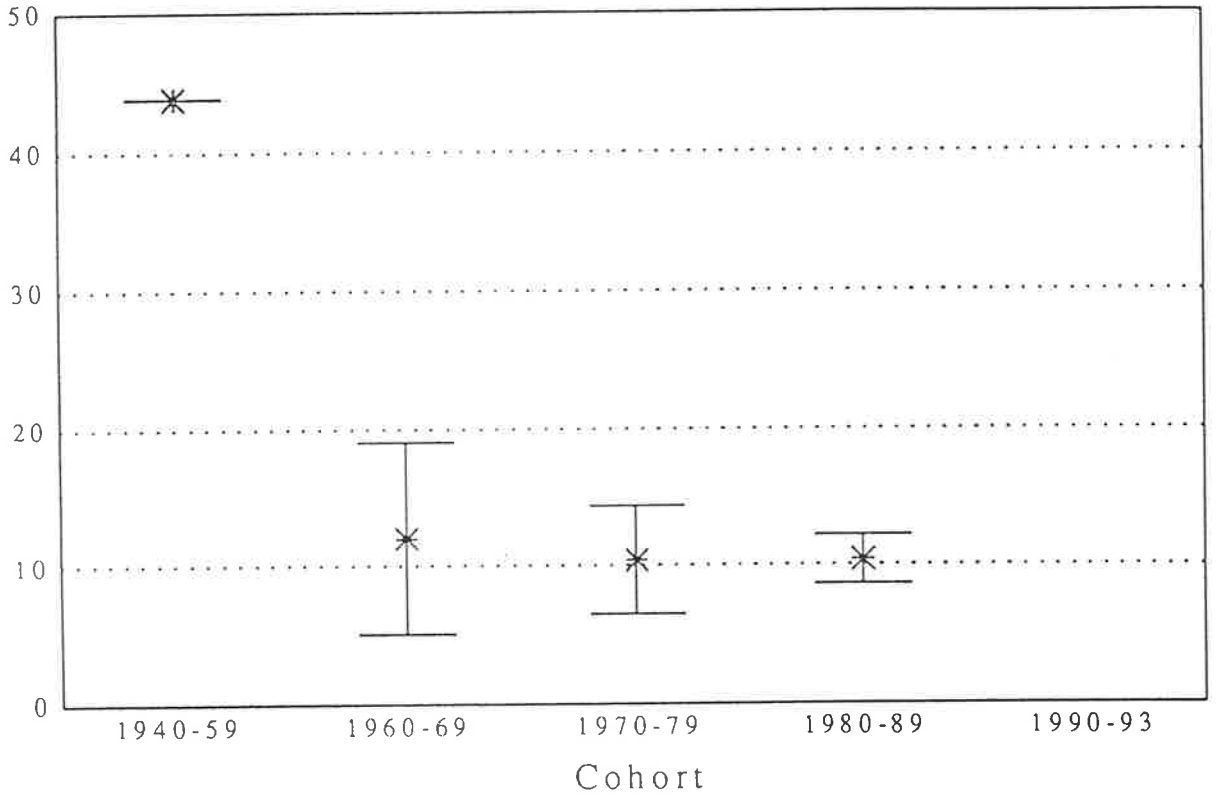


Figure 3.7: Mean age of intervention - Pharyngoplasty

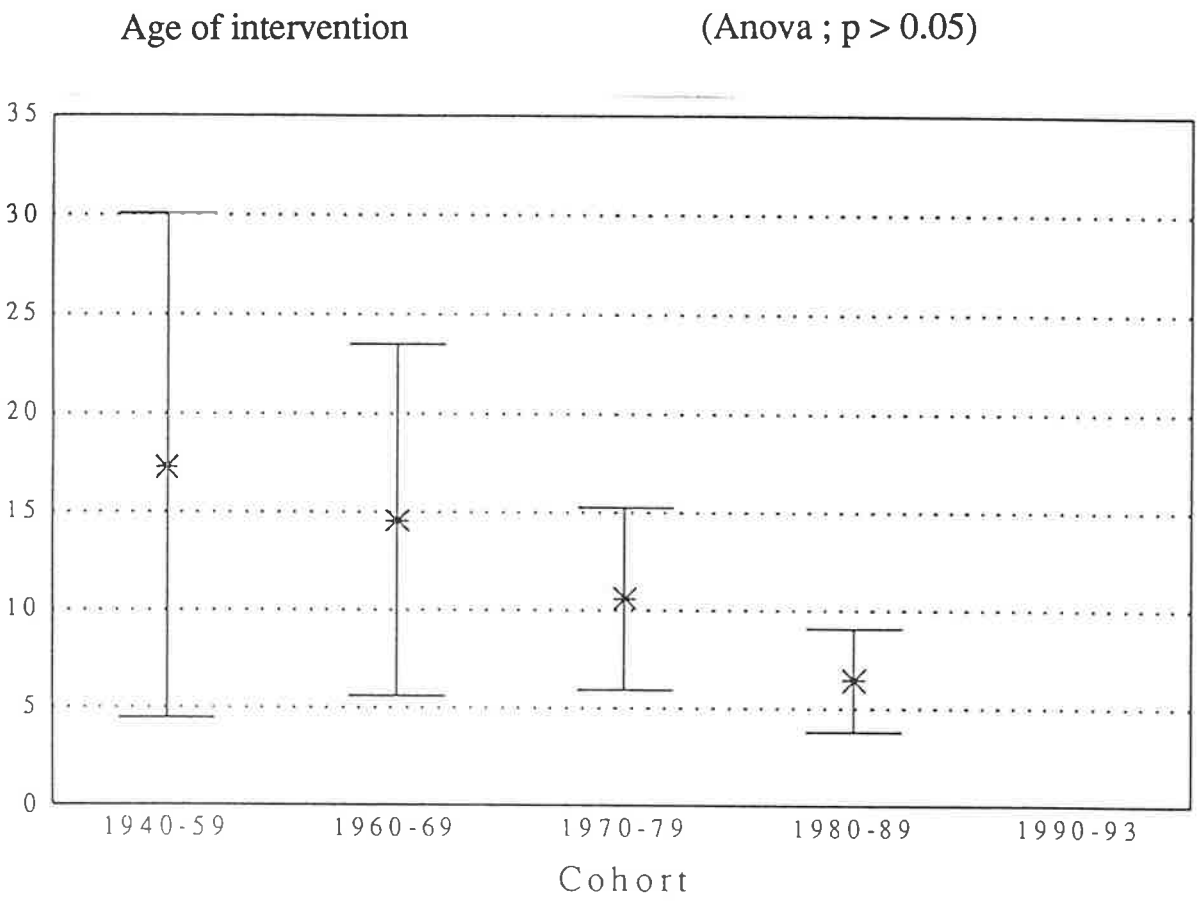
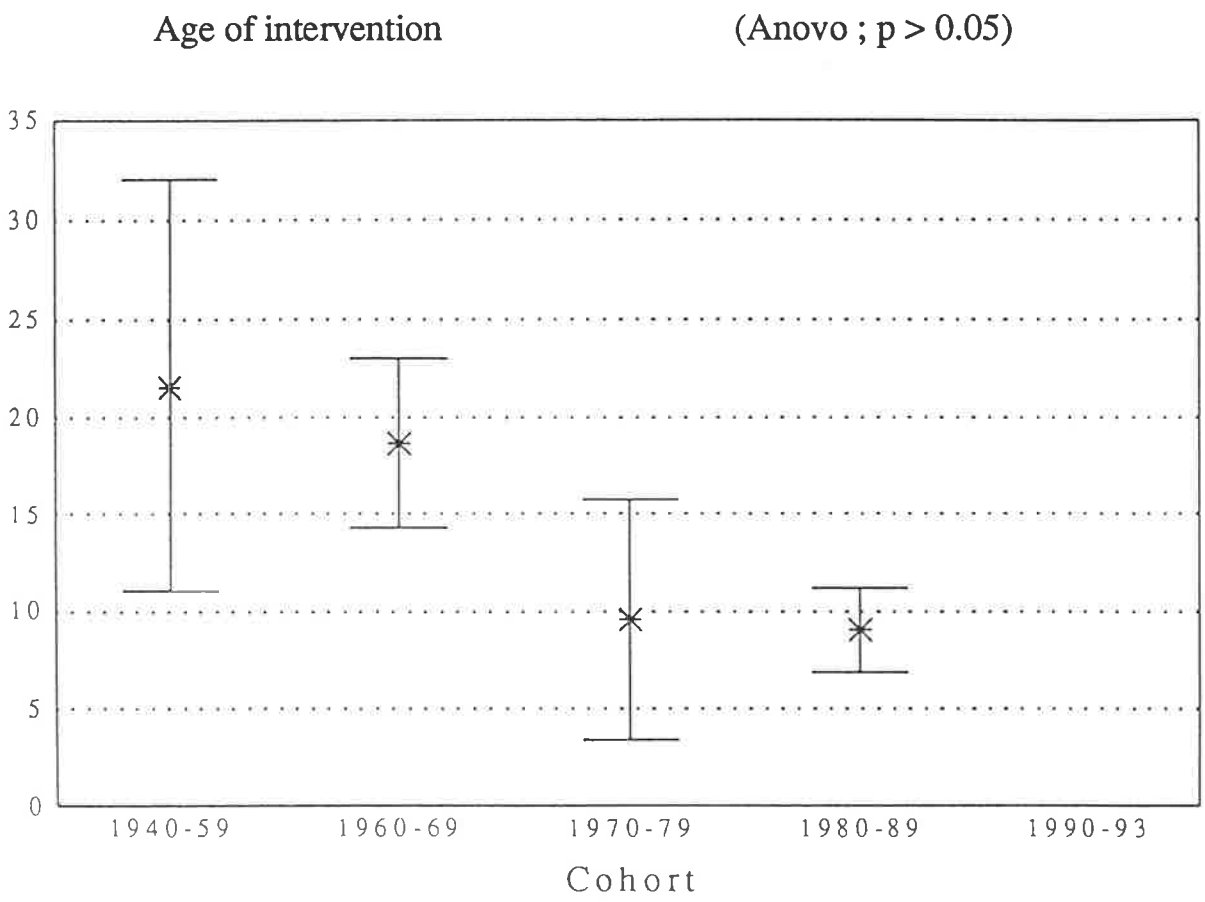


Figure 3.8: Mean age of intervention - Rhinoplasty



BILATERAL MYRINGOTOMIES

From Figure 3.9, it can be observed that that mean age of intervention has been decreasing over last five decades. The deviation from the mean has also been decreasing in the last five decades. By the years 1990-93 the age of intervention is between 1 - 3 years (Appendix C6).

OSTEOTOMIES

From Figure 3.10, it can be observed that the mean age of intervention for osteotomies has decreased. The deviation decreased until 1980, but then increased slightly from 1980 onwards. Most of the osteotomies by the period of 1993 were done by the age of 13 - 16 years (Appendix C7).

ORTHODONTICS

As seen in Figure 3.11, the mean age of intervention for orthodontic treatment has rapidly decreased since the 1960s birth cohort. The standard deviation has also decreased across the last threecohorts. By the 1990s cohort most of the interventions were carried out at the age of 5 -11 years (Appendix C8).

Figure 3.9: Mean age of intervention - Bilateral myringotomies

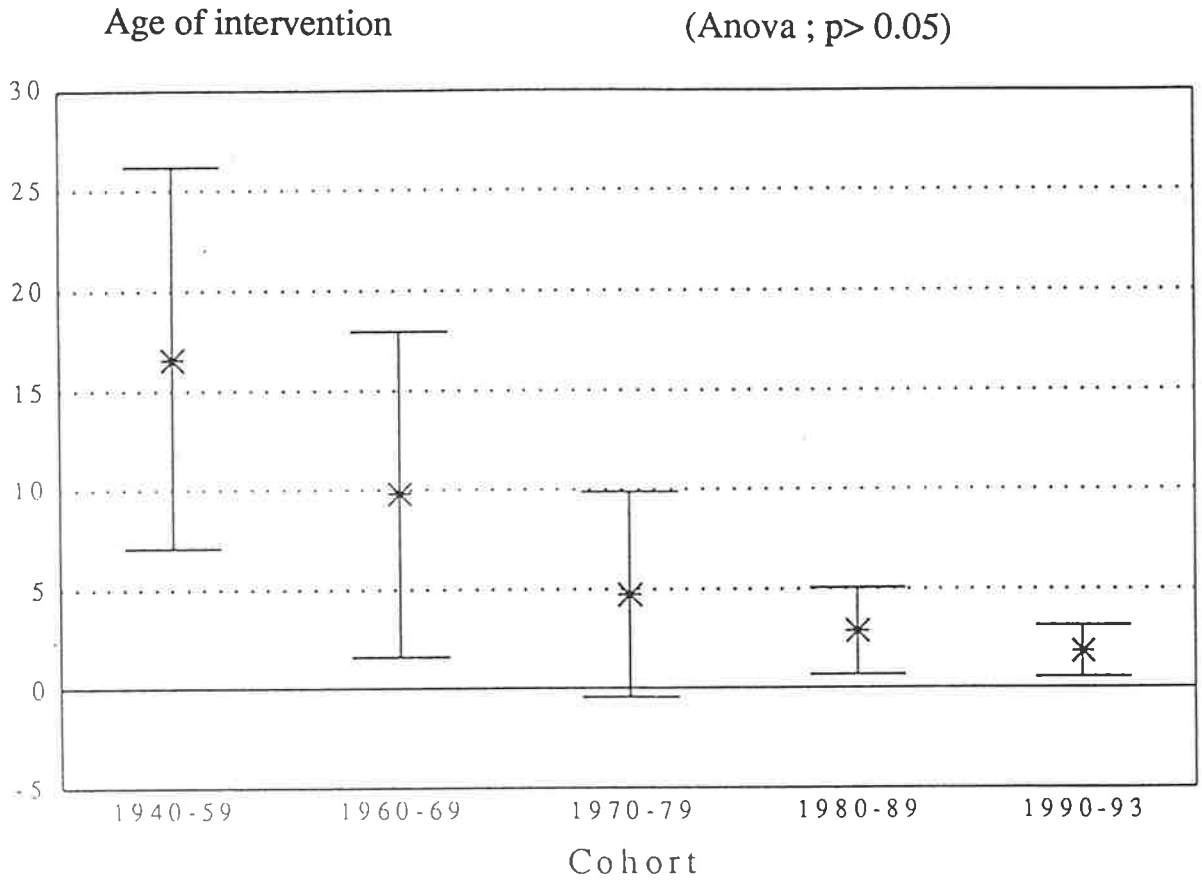


Figure 3.10: Mean age of first intervention - Osteotomy

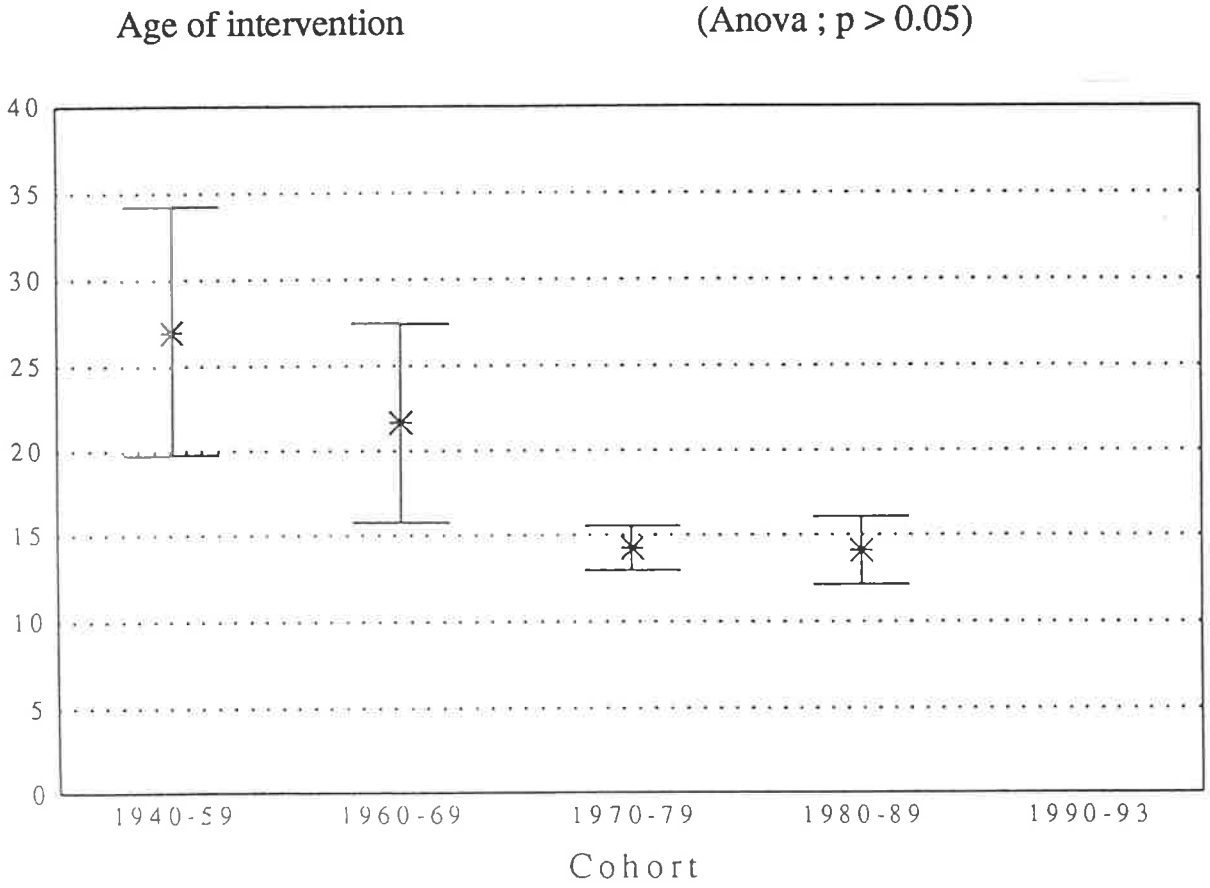
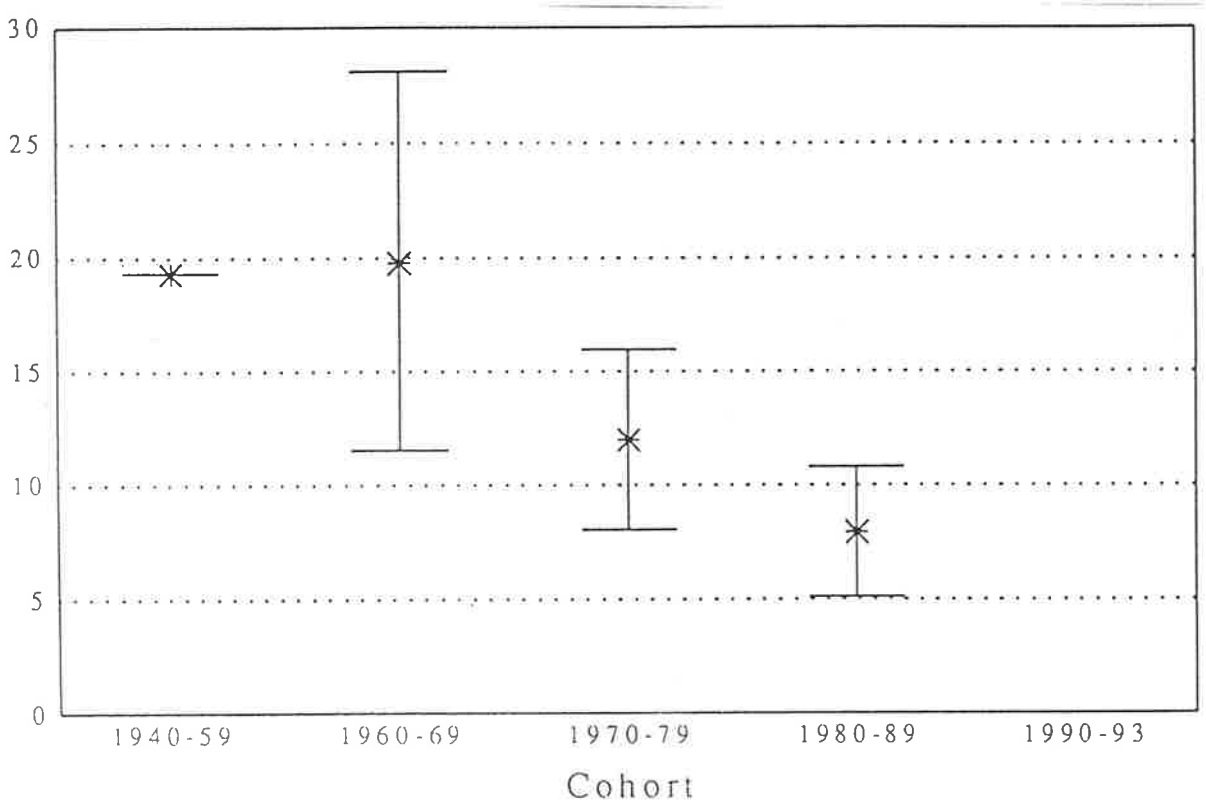


Figure 3.11: **Mean age of intervention - Orthodontics**

Age of intervention (Anova ; $p < 0.05$)

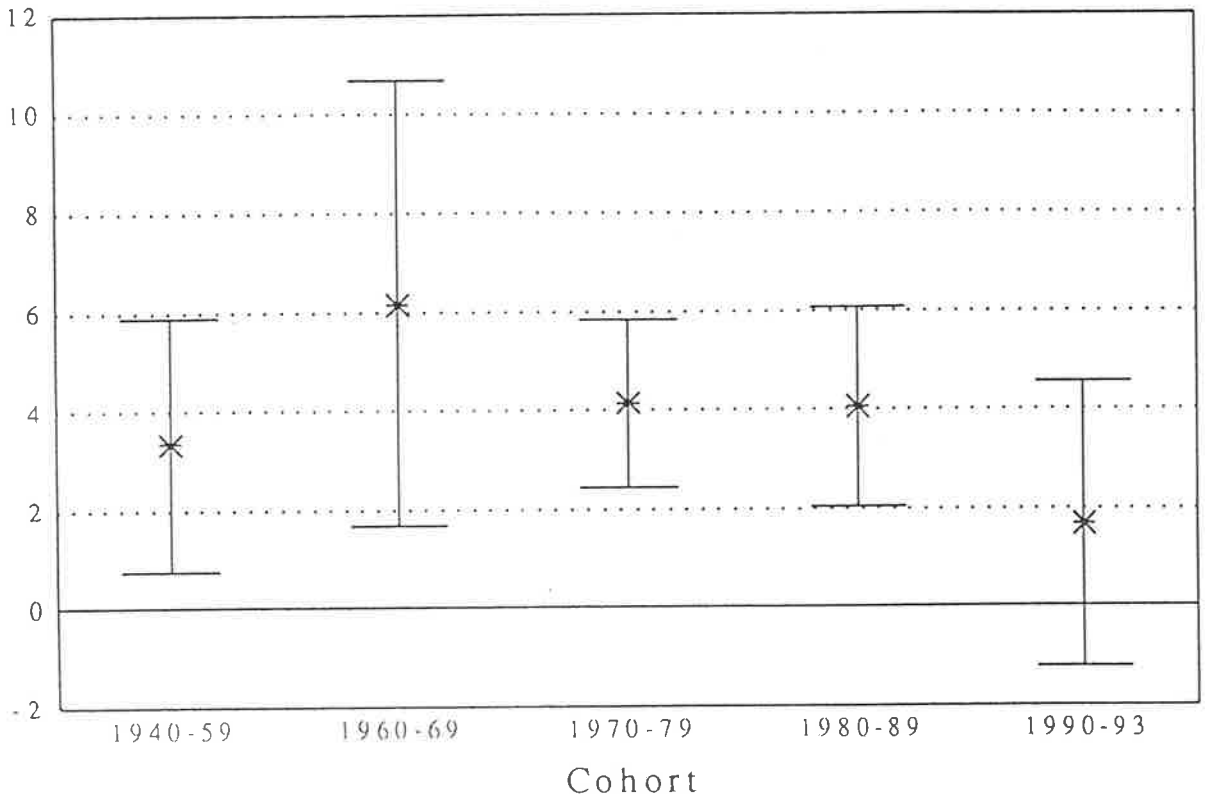


SPEECH THERAPY

Figure 3.12 illustrates the mean age of intervention for speech therapy. The mean has not been very consistent over the birth cohorts. There was an increase in mean age to the 1960s cohort, then a decrease for the 1970s and 1980s cohort. This decrease was more pronounced for the 1990s birth cohort, where the mean age of intervention has decreased considerably. However, for 1990s birth cohort the standard deviation has significantly increased. The interventions has been carried out at a range of ages, from less than one year to five years of age (Appendix C9).

Figure 3.12: Mean age of intervention - Speech therapy

Age of intervention (Anova; $p < 0.05$)



3.2.2 DISTRIBUTION OF AGE OF FIRST INTERVENTION

This analysis was done to find out at which particular age did the majority of the sample have their first surgical intervention. It was recognised that the distribution of age of first surgical intervention might be non - symmetrical, leading to the mean being misleading with regard to the age of first intervention.

REPAIR OF CLEFT LIP

The vast major of the sample, 87.9 %, had their repair of cleft lip at the age of 3-4 months. A small minority of the sample, 6% had the repair at the age of one year, and the subjects are seen to have the repair done at various other ages. The details are seen in Figure 3.13 (Appendix D1).

REPAIR OF CLEFT PALATE

The age of first intervention for repair of cleft lip was between 6-12 months for 74.9 % of the study samples. About 15% had repair of cleft palate at two years of age. And rest of the sample had the repair of cleft palate at various other ages. The frequency distribution is seen in Figure 3.14 (Appendix D2).

Figure 3.13: Age of first intervention - Repair of cleft lip

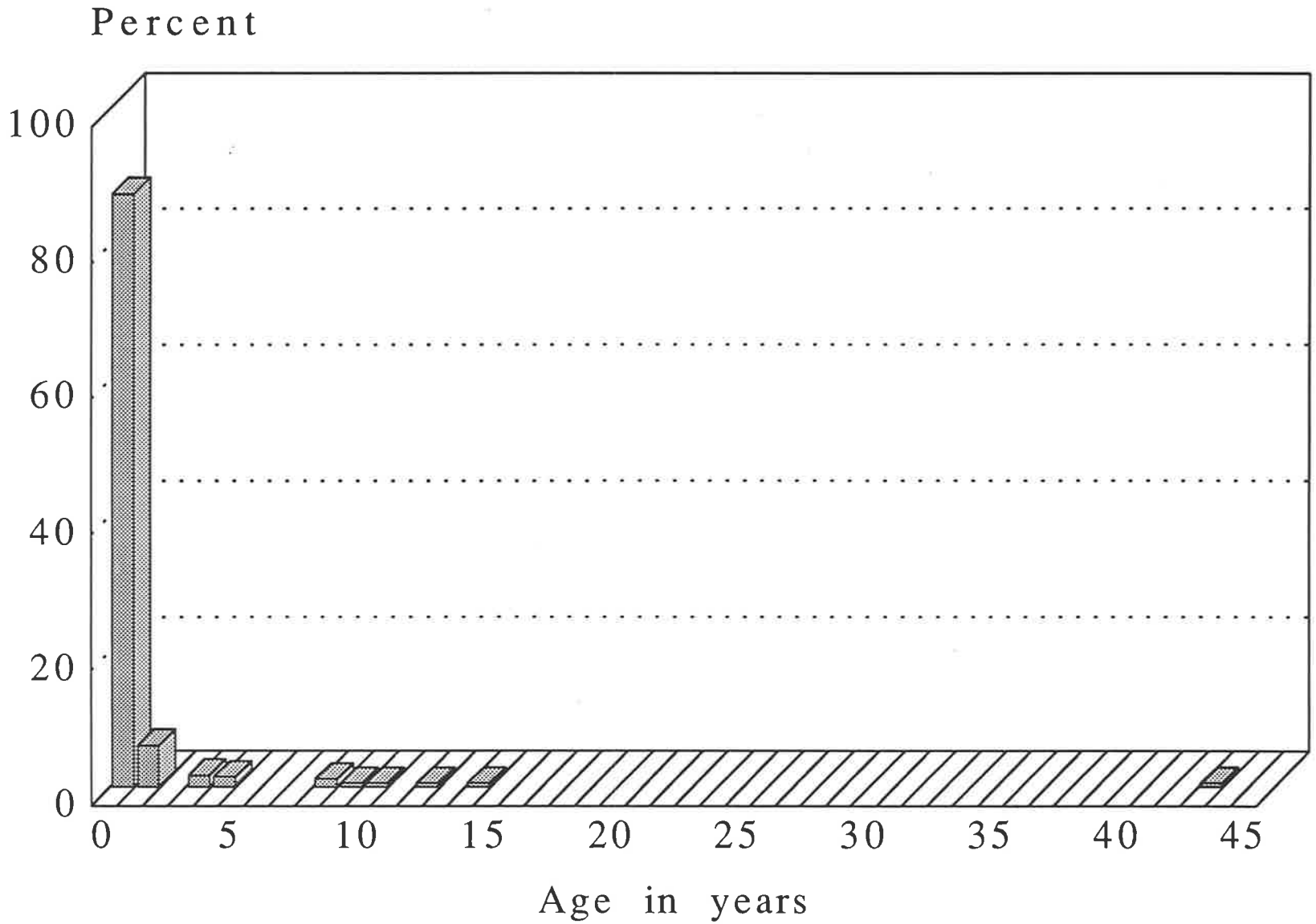
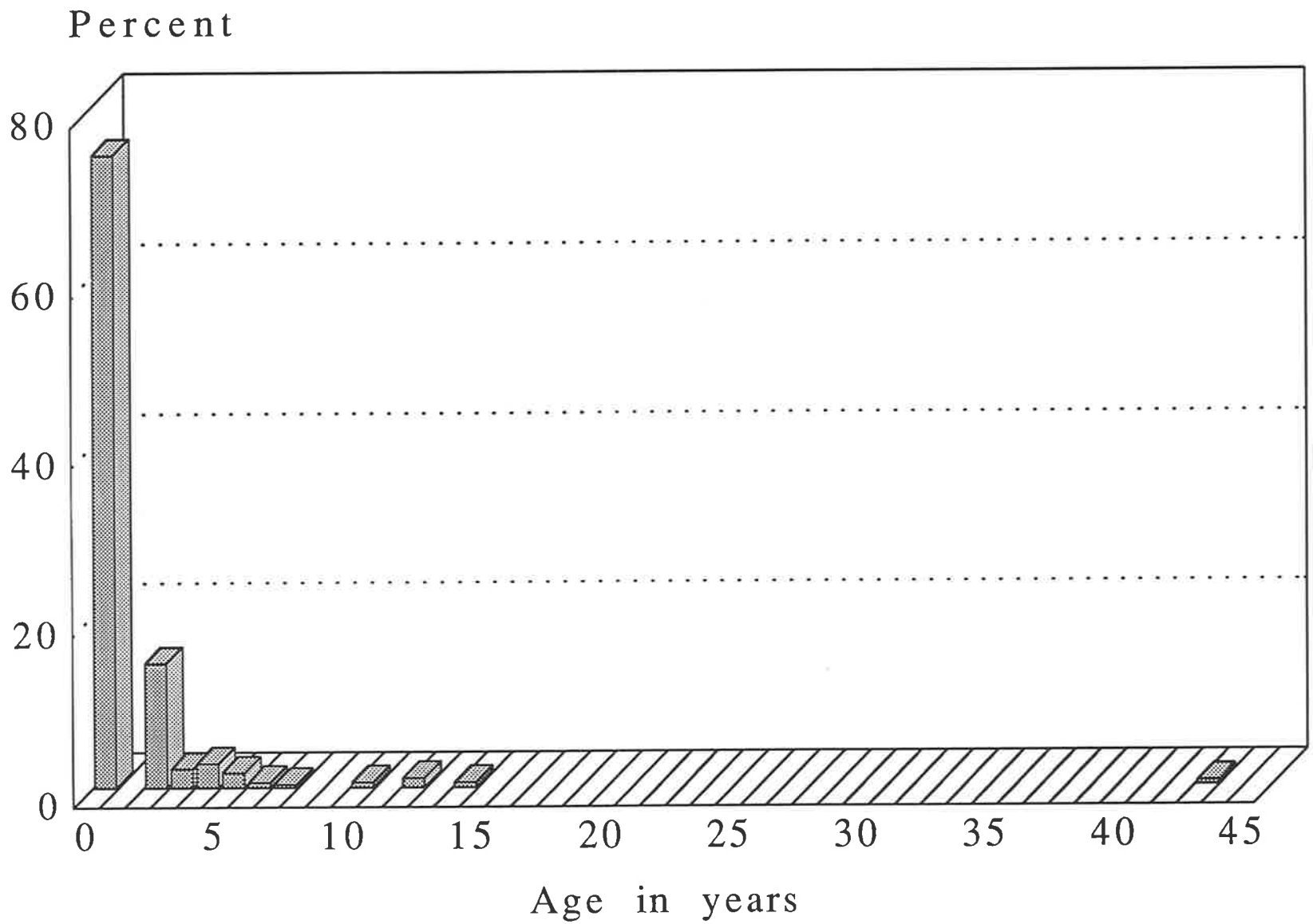


Figure 3.14: Age of first intervention - Repair of cleft palate



AVEOLAR BONE GRAFTING

Referring to Figure 3.15, about 32% of the study sample had their alveolar bone grafting at 12 years of age and other substaintial percentages of the subjects had this intervention done at the age of nine years and 14 years, 14.3 % and 11.5 % respectively (Appendix D3).

PHARYNGOPLASTY

26 percent of the study population had their pharyngoplasty at four years of age. Another increased percentage is seen at seven years of age where 21.5 percent of the study population had pharyngoplasty. Most of the other subject cases are distributed in other ages. Figure 3.16 illustrates the frequency distribution. (Appendix D4)

NASAL TIP REVISION / RHINOPLASTY

As seen in Figure 3.17, 34.8 percent of the study population had their first intervention for rhinoplasty done at the age of 12 years. Followed by 19.5 percent at 14 years and 10.9 percent at 16 years of age. There was a small distribution of study population at other ages. (Appendix D5)

Figure 3.15: Age of first intervention - Alveolar bone grafting

Figure 3.16: Age of first intervention - Pharyngoplasty

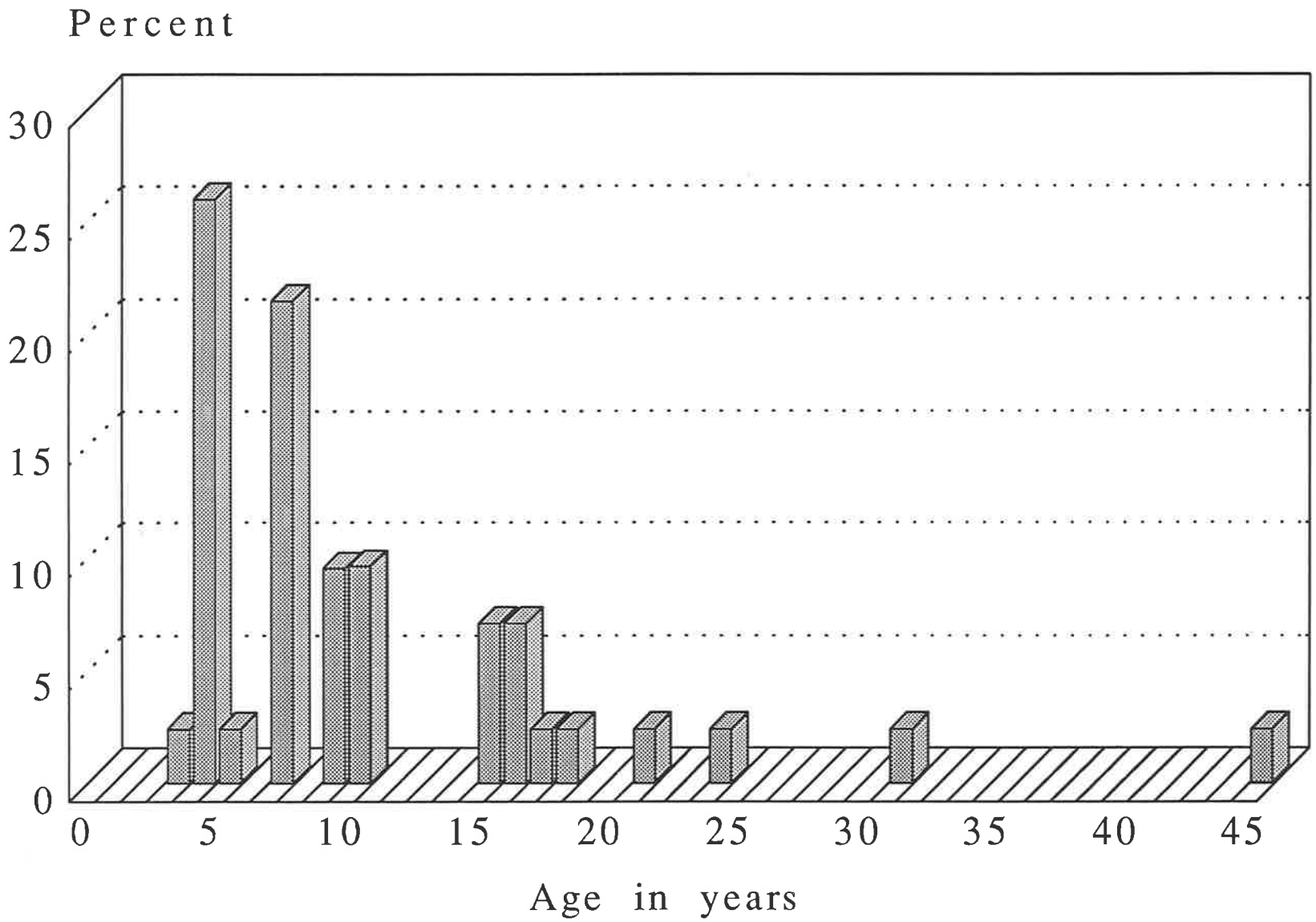
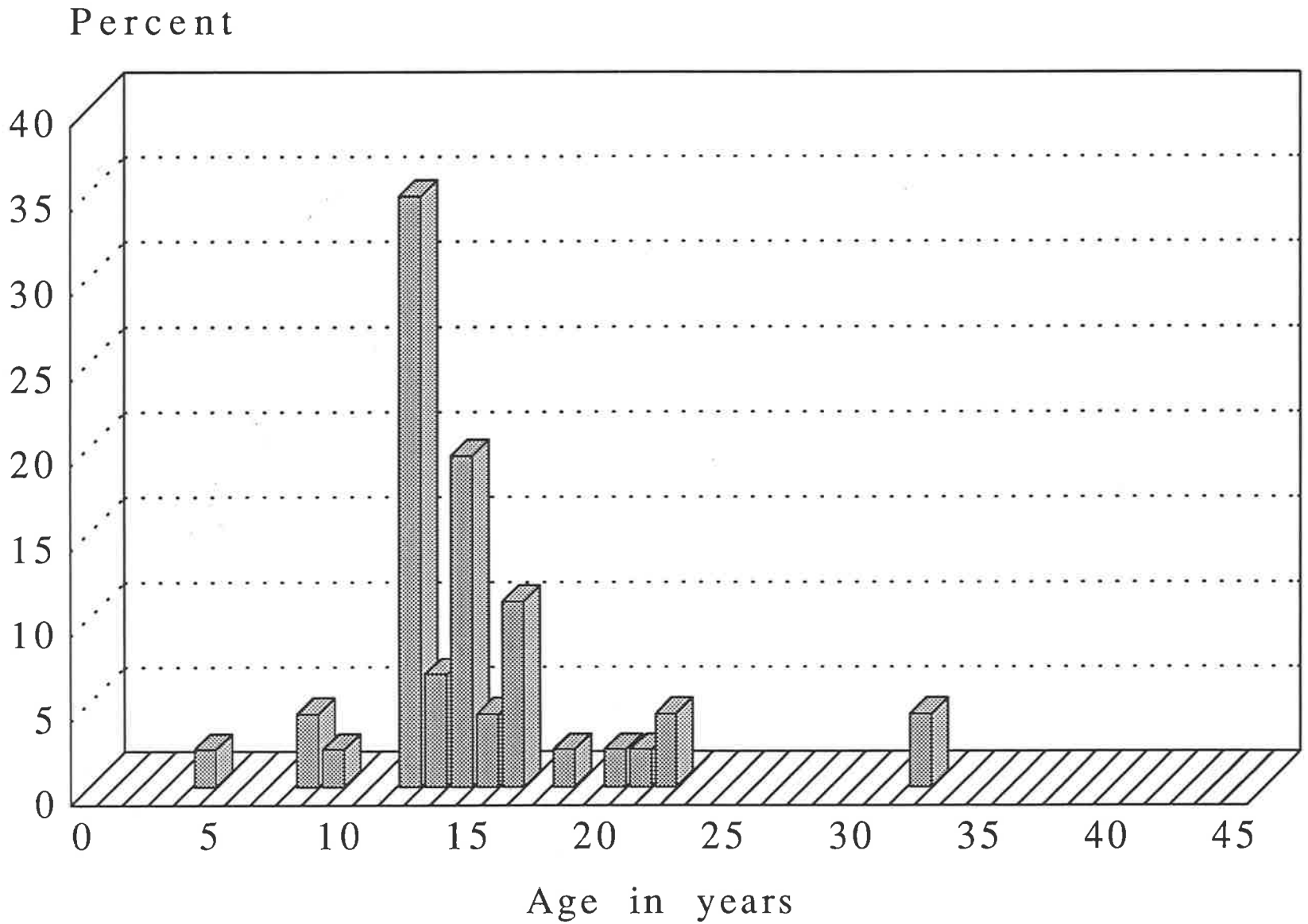


Figure 3.17: Age of first intervention - Rhinoplasty / Nasal tip revision



BILATERAL MYRINGOTOMIES

Most of the cases had their first insertion of ventilation tubes in their ears in the age ranging from one year to five years. 27.4 percent at two years followed by 21.1 percent at three years and 19 percent at one year of age. The remaining cases in the study population are distributed in various older ages. Figure 3.18 illustrates the frequency distribution. (Appendix D6)

OSTEOTOMIES

Of the study population 44 percent had cranio maxillary facial surgery at 15 years of age. 22 percent had the surgery done at 16 years of age. There were low percentages of the cases who had their osteotomies done at 12,14, and 19 to 32 years of age.

Figure 3.19 illustrates the frequency distribution. (Appendix D7)

Figure 3.18: Age of first intervention - Bilateral Myringotomies

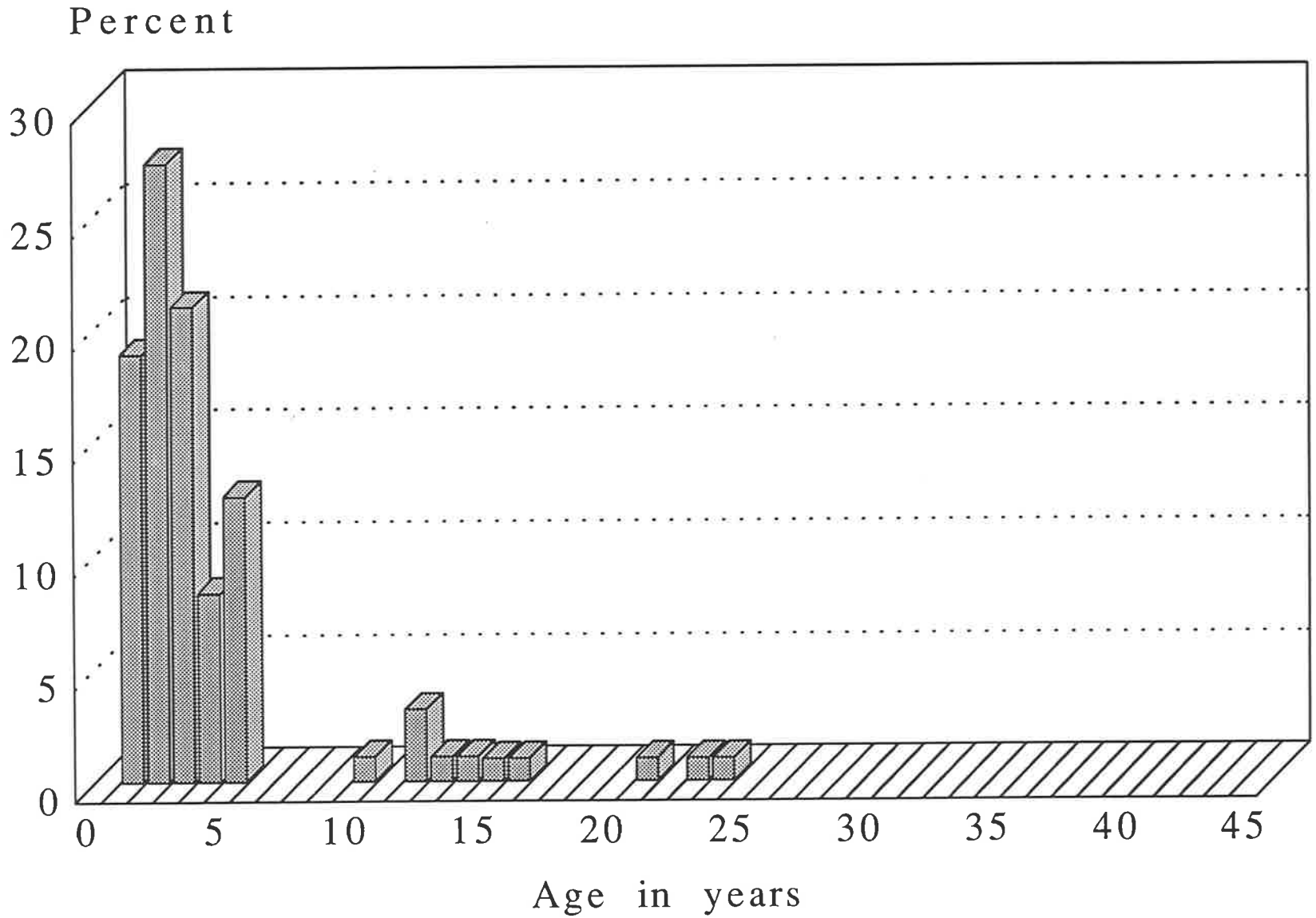


Figure 3.19: Age of first intervention - Osteotomies

3.2.3 POPULATION PERCENTAGE AND NUMBER OF DAYS IN THE HOSPITAL

This analysis was done to estimate the number of days the patient spent in the hospital from the day of admission to the day of discharge, which generally is after cranio maxillary facial surgery or osteotomies. As seen in the Figure 21, the time ranged to minimum of six days to maximum of 50 days in the hospital in account of surgical intervention for correction of cleft defects. Other time spent in the hospital due any other medical condition or surgical conditions was not recorded. It is found that 31 percent of the study population spent about 20 days in the hospital, about 20 percent of the population spent 10 days and rest of the frequencies can be appreciated in figure 3.20. (Appendix E)

Figure 3.20: Number of days in the hospital for various surgical interventions related to correction of facial clefts

3.2.4 MEAN AGE OF TREATMENT FOR VARIOUS SURGICAL PROCEDURES BY COHORT FOR GROUP I, GROUP II AND GROUP III CLEFTS

The classification for clefts is based on International Classification for Cleft lip, Alveolus and Palate (Hall, 1994). This classification was used to examine variation in the treatment by cleft classification. The principal features of the treatment and mean age of treatment are indicated in Table 3.1 for Group I , Table 3.2 for Group II and Table 3.3 for Group III. The mean and standard deviation for each interventionare presented.

Summary of treatment for Group I cleft by cohort

The main surgical interventions for patients in Group I Clefts were primary repair of cleft lip, rhinoplasty / nasal tip revision.

Table 3.1 shows the mean age and deviation for the surgical interventions for Group I clefts by birth cohort.

TABLE 3.1 : Mean age of various surgical interventions by cohort for Group I clefts

COHORT	Repair of Cleft Lip*		Rhinoplasty/Septoplasty	
	Mean	S.D	Mean	S.D
1960 -1969	4.7 yr	7.5 yr	21.14 yr	0.00 yr
1970 - 1979	1.4 yr	2.8 yr	18.3 yr	0.77 yr
1980 - 1989	0.44 yr	0.1 yr	16.8 yr	0.07 yr
1990 - 1993	0.39 yr	0.018 yr	14.05 yr	0.7 yr

S.D = STANDARD DEVIATION
* Anova ; $p < 0.05$

Table 3.1 shows that by the 1980s cohort there was a considerable amount of stability in the timing of the repair of cleft lip as well as rhinoplasty. Both interventions also had reduced standard deviation with time . During 1960-1969 the repair of cleft lip occurred between 4.7 ± 7.5 years of age and rhinoplasty was done at 21.14 years of age approximately.

Among the cohort born between 1970 to 1979 the mean age and standard deviation for both repair of cleft lip and rhinoplasty decreased. 1980 onwards the repair of cleft lip was done between three to five months of age and rhinoplasty was done at 13 to 17 years of age.

Summary of treatment for Group II cleft by cohort

The main interventions for patients in Group II clefts are primary lip repair, repair of cleft palate, alveolar bone grafting, rhinoplasty, pharyngoplasty, osteotomies and bilateral myringotomies.

Table 3.2 shows the mean age and deviation for various interventions for Group II clefts by cohort. The cohort born between 1990 to 1993 is not included as treatment has not been completed for these subjects.

TABLE 3.2 : Mean age of various surgical interventions by cohort for Group II clefts

SURGICAL INTERVENTIONS	COHORT			
	1940-1959	1960-1969	1970-1979	1980-1989
Repair of Cleft Lip*	0.3 ± 0.3 yr	3.6 ± 3.4 yr	0.37 ± 0.55 yr	0.38 ± 0.64 yr
Repair of Cleft Palate*	1.2 ± 0.4 yr	2.2 ± 3.4 yr	1.3 ± 0.4 yr	1.2 ± 1.1 yr
Alveolar Bone Graft*	16 ± 1.2 yr	12 ± 7 yr	0.5 ± 3.8 yr	10.2 ± 2.1 yr
Rhinoplasty*	21.9 ± 12.15 yr	18 ± 4.8 yr	9.1 ± 6.3 yr	8.4 ± 2.2 yr
Pharyngoplasty*	18.5 ± 10.9 yr	14.5 ± 9 yr	11.6 ± 4.1 yr	4.2 ± 2.7 yr
Osteotomies	25.4 ± 7.3 yr	21.5 ± 5.8 yr	14.5 ± 1.1 yr	13.3 ± 1.1 yr
Bilateral Myringotomies	10.3 ± 0.0 yr	9.7 ± 8.2 yr	4.7 ± 5.7 yr	2.6 ± 1.8 yr

* Anova ; $p < 0.05$

As seen in the above table (Table 3.2), the mean age for repair of cleft lip was similar in all cohorts being between 3 - 4 months except in cohort born between 1960 to 1969 where the mean age for the repair of lip was 3.6 years.

The mean age of repair of cleft palate was also very similar in all cohorts being 1.2 years to 1.3 years except in the cohort born between 1960 to

1969 where the intervention was done at a mean age of 2.2 years. There was a continuous decrease in the mean age of intervention for alveolar bone grafting, the mean age being 16 years in 1940 to 59, 12 years in 1960 to 1969, 10.5 years in 1970 to 1979 and 10.2 years 1980 onwards. The mean age for rhinoplasty also showed a continuous decrease, from being 21.9 years in cohort born between 1940 to 1959, 18 years for the cohort born between 1960 to 1969, dropping to 9 years for the cohort between 1970 to 1979 and 8.4 years for the cohort born from 1980 onwards. A similar trend is seen in pharyngoplasty, the mean age being 18.5 years for the cohort born between 1940 to 1959, 14.5 years for the cohort born between 1960 to 1969, 11.6 years for the cohort born between 1970 to 1979 and a drop to 4.2 years for the cohort born between 1980's.
The mean age for osteotomies has also reduced over time. The mean age was 25.4 years for the cohort born between 1940 to 1959 and since then gradually decreasing to reach a mean age of 13.3 years for the cohort born between 1980s. The mean age for bilateral myringotomies for the cohort born between 1940 to 1959 was 10.3 years, but among the cohort born in 1980 onwards the surgery was done at a mean age of 2.6 years.

These data showed that there was a continuous drop in the mean age of most of the interventions with time and the standard deviation has also decreased for most of the interventions. Most of the interventions are

carried out at a particular time interval for most of the patients and thereby completing the complete treatment at a much earlier age.

Summary of treatment for Group III clefts by cohort

The main surgical interventions involved in correction of Group III clefts are repair of cleft palate, pharyngoplasty and bilateral myringotomies. The principal features of the treatment and the mean age for the interventions are described in Table 3.3.

TABLE 3.3 : Mean of various surgical interventions by cohort for Group III clefts

SURGICAL INTERVENTIONS 1990	COHORT				
	1940 1959	1960 1969	1970 1979	1980 1989	1993
Repair of Cleft Palate*	6.5 ± 7.6 yr	---------	3.3 ± 3.9 yr	1.8 ± 2.1 yr	0.63 ± 0.0 yr
Pharyngoplasty	14.7 ± 0.0 yr	---------	7.5 ± 3.5 yr	3.2 ± 2 yr	2.8 ± 0.09 yr
Bilateral Myringotomies*	9.8 ± 0.0 yr	--------	4.6 ± 3 yr	3.1 ± 2.6 yr	1.5 ± 0.0 yr

* Anova ; $p < 0.05$

As seen in the Table 3.3, the mean age for all the three intervention has reduced over time. The standard deviation and the mean has also

considerably reduced. The surgical treatment is completed much earlier in life since the 1980s than what it use to be in earlier times.

3.3.5 COMPARISON OF CLEFT CLASS BY AGE OF INTERVENTIONS FOR COHORT 1980-1989

Comparison of mean age of various intervention by cleft classification is done for subjects born in cohort 1980-1989 as most of the study subjects belonged to this cohort and enough data could be collected to make comparisons for mean age for various intervention by cleft class.
The cohort 1960-1969, and 1970-1979 are not used in the comparison as no statistically significant data could not be extracted for various intervention for correction of cleft lip/ or palate.

The main interventions used in comparison of mean age by cleft class are repair of cleft lip, repair of cleft palate, alveolar bone graft, rhinoplasty /or nasal tip revision and pharyngoplasty. Table 3.4 shows the mean age of intervention by cleft class.

Table 3.4: Comparison of cleft class by age for cohort 1980-1989

SURGICAL INTERVENTIONS	CLASS I	CLASS II	CLASS III
Repair of Cleft Lip	0.44 ± 0.1	0.38 ± 0.64	-------
Repair of Cleft Palate	--------	1.2 ± 1.1	3.1 ± 2.6
Alveolar Bone Graft	8.77 ± 1.01	10.2 ± 2.1	-------
Rhinoplasty	16.8 ± 0.07	8.4 ± 2.2	-------
Pharyngoplasty	-------	4.2 ± 2.7	3.2 ± 2.

As seen in above Table 3.4 Repair of lip was done at a mean age of 0.44 months for class I clefts and at 0.38 months for class II clefts and their is a decrease in standard deviation for class I clefts.

Repair of cleft palate for class II clefts was carried out at a mean age of 1.2 years with a standard deviation of 1.1 and for class III clefts was carried out at 3.1 years and with an increased standard deviation of 2.6. Their is difference in timing of

repair of cleft palate for class II and Class III this may be due to various other surgical interventions involved in class II clefts and the severity of the defect. Alveolar bone grafting was carried out an mean age of 8.77 years for class I clefts and at 10.2 years for class II clefts. Standard deviation is seen to be increased in class II clefts. This also may be due to involvement of various other surgical interventions involved in correction of class II cleft.

The mean age for rhinoplasty or nasal tip revision for class I cleft is 16.8 years with a standard deviation of 0.07 months, the mean age for class II cleft is 8.4 years and standard deviation is 2.2. The difference in mean age for rhinoplasty or nasal tip revision for class I and class II may be due to difficulty in extracting the data from the subject records due to unavailability of proper information of the intervention ie, whether it was rhinoplasty or repair of nasal tip.

Pharyngoplasty was carried out at an mean age of 4.2 years for class II clefts and at 3.2 years for Class III cleft and with a standard deviation of 2.7 for class II and 2 for class III.

4: DISCUSSIONS

4.1 OVERVIEW

The obvious variation in the age at which various surgical, medical and dental interventions were carried out between the birth cohorts leads to some interesting conclusions. The mean scores for all the intervention carried out in treatment of cleft lip and / or palate in all the cohorts were significantly different. The mean age value were higher in first birth cohort and reduced with each consecutive cohort. The deviation also was large in the first birth cohort. This indicates that there was no standardised policy followed for various interventions for the earlier birth cohorts, and the interventions were frequently carried out late in the child's development. The considerable drop in the mean age for surgical interventions for the 1980s birth cohort and the 1990s birth cohort and decrease in the standard deviation around the mean are suggestive of :

1. a long term evaluation of the surgical interventions;
2. the interventions being based on scientific evaluation of craniofacial growth;
3. a multidisciplinary approach enabling evaluation and planning of a proper treatment strategy;
4. the following of standardised treatment policy in recent times; and
5. a reduced rate of failure of the particular intervention.

There is a more standardised treatment policy, and the patients complete their treatment much earlier in life due to an improved treatment approach and planning.

The improved treatment approach should lead to reduced rates of surgical failure and secondary interventions and better results aesthetically, functionally and socially. Early speech intervention and speech therapy at regular intervals gives the child a better chance to lead a normal social life.

The considerable change in the treatment profile over the years and standardised policy with regard to age of intervention strongly supports the view that the team approach is the solution for better management of cleft lip and / or palate, leading to more desirable results with less failures and a decreased total period of hospitalisation for these cases.

4.2: MANAGEMENT OF CLEFT LIP AND PALATE AT THE AUSTRALIAN CRANIO FACIAL UNIT OVER LAST 15 YEARS

The management of cleft lip and / palate over last 15 years at the Australian Cranio Facial Unit based at Adelaide Children's Hospital includes a multidisciplinary team involved in interventions for cleft lip and / or palate, the timing of which extends from birth until the completion of full development ie, during early adulthood.

The Australian Cranio Facial Unit team composes of :

Plastic Surgeon;

Paediatrician;

Speech Pathologist;

ENT Surgeon;

Orthodontist;

Genetic Counsellor; and,

Pedodontist.

The main aim of the Unit is that of a centralised team and treatment over period of time, leading to a normal development of the patient's face,

provision of emotional support to the families, and facilitation of parent / professional collaboration in the health care of children.

To evaluate the child and to plan a proper treatment strategy, a team meeting is organised which is composed of various members. Figure 4.1 shows all the components of the Australian Cranio Facial Unit team.

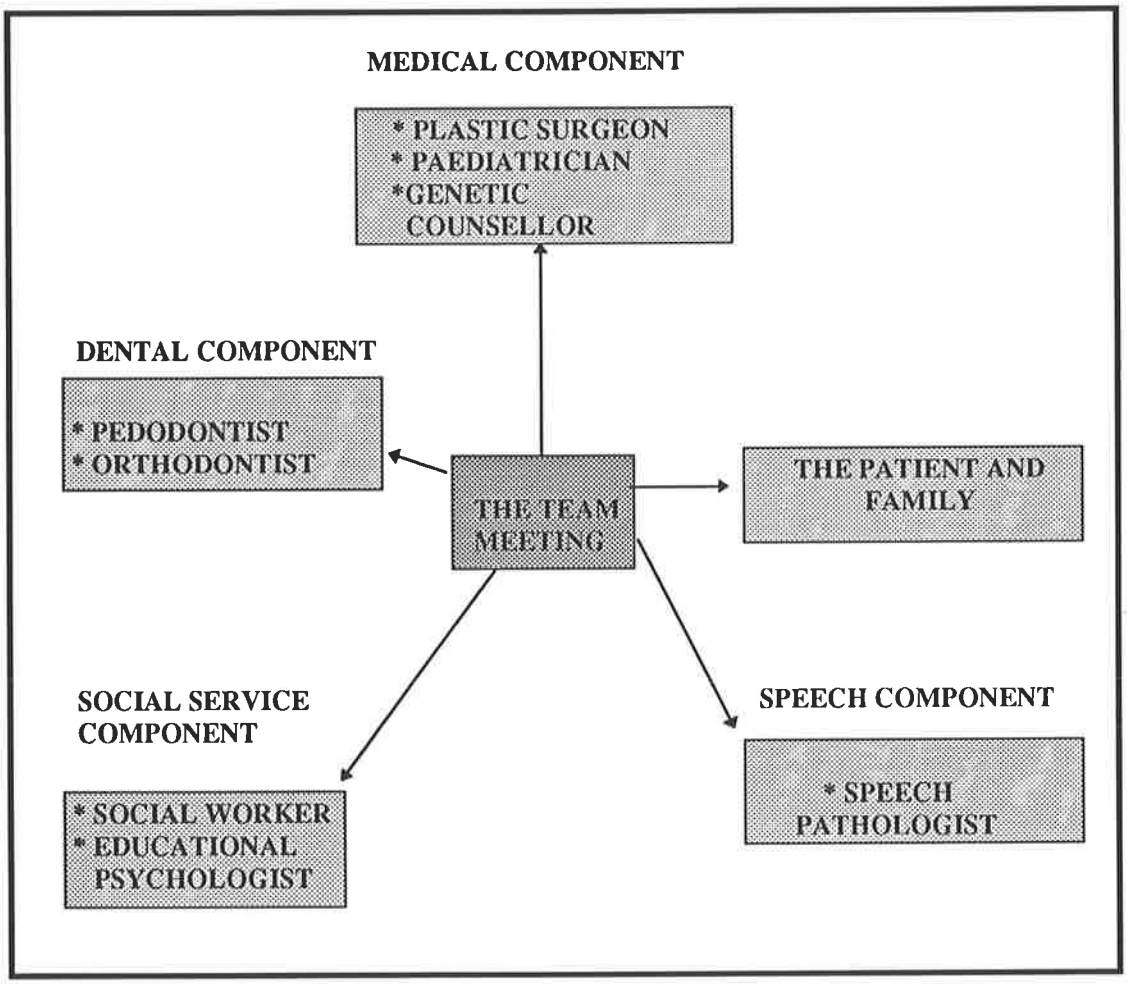


FIGURE 4.1: Components of cranio facial team meetings

On reviewing the results derived from the study of cases born after 1980, the following protocol for treatment of cleft lip and palate can be inferred (based on Lecture by David. J. David, 1994):

Management of the patient begins the day he or she is born. It begins on day one with the initial visit with counselling by the surgeon and advice on early feeding is done by the speech pathologist.

Repair of cleft lip is done at 3 - 4 months of age and stiches are removed seven days following surgery. Review by the surgeon is done4 - 6 weeks following surgery. Photographs are taken before and after surgery.

Repair of cleft palate is done from 6 - 12 months of age. Review by surgeon is done 4 - 6 weeks following surgery. Photographs are taken before and after surgery.

Between the age 6 - 8 months an E.N.T consultation is done to asses the status of eustachian tube function and middle ear function and if necessary insertion of ventilation tubes (**bilateral myringotomy**) is done at an age of two years.

Speech assessment begins at one year and five months of age and yearly evaluation is done. Along with speech assessment at about the age of 4 - 6 years of age nasendoscopy is done.

Depending on nasendoscopy results, if **pharyngoplasty** is indicated it is carried out between 4 - 6 years of age.

Orthodontic assessment is done at 7 - 12 years of age. Radiographs are taken, eg, orthopantamogram, lateral cephalogram, hand wrist, etc. Cephalometrix X-rays are then taken every year until growth and treatment is completed.

After the orthodontic assessment **alveolar bone grafting** is done between 10 - 12 years of age.

At about 12 -13 years of age maxillo-facial orthodontic assessment is done to find weather patient fits into the category of orthodontics alone or requires a surgical procedure in addition to orthodontic treatment. Depending on the assessment **osteotomies** are done between 13 -15 years of age.

Rhinoplasty is done between 9 -15 years of age. It can be done at the same time when the osteotomy is performed.

Genetic counselling, psychosocial assessment, dental assessment, photographs, radiographs and treatment are regularly done throughout the growth period and until the treatment is complete.

The Cranio Facial Unit team meetings along with the patient and parents is done on yearly basis after the assessment of cranio facial growth and development with the orthodontist. And these meetings are organised with the patient and the parents consisting of a complete team assessment involving all appropriate disciplines until all the treatment is complete. The team employs biplanar cephalometric x rays and more sophisticated CAT scans which gives the team an opportunity to study and evaluate the real three dimensional nature of clefts and helps the team to focus on the proper three dimensional correction of the deformities. Post surgical follow up is done during the team meetings to monitor the results and relapse rate. If any touch up or secondary surgery are required, they are carried out in conjunction with other surgeries or a suitable time for the surgery is decided. This reduces hospitalisation, which these patients face frequently due various surgeries involved in correction of these clefts. This also helps

the team to complete all the treatment required by the time patient reaches early adulthood.

4.3 STUDY LIMITATIONS

With an incidence around one birth per six hundred births, and the lack of detail in hospital records, a study sample of 255 cases were only available for this study. However, the sample served to be adequate for hypothesis testing across birth cohorts. There was a considerable variation in sizes between birth cohorts. This is due to lack of information within the medical records, or the case notes not being available or the records had been microfilmed, or discontinuity of treatment by the patient due to change of State or country.

Descriptive cleft classification was not available in most of the case notes which prevented the description of anatomical extent of the cleft, hence those subject were not incorporated in the study. The dates of interventions could be extracted from the medical records, but what procedure or technique was used for the interventions could not be extracted from many of the case notes of the study population.

Due to all the above reasons there is variation in sample size between cohorts . It is recognised that the variable sample sizes will have some influence on the statistical power of comparisons between cohorts. Beyond this refinement of individual techniques or variations of the surgical program can only be evaluated and advanced through prospective trials, preferably on multi center basis. Single center

study designs do not lend themselves to efficacy studies because of difficulties in building to sufficient subjects, but they serve well to generate hypotheses for more rigorous research and testing.

5: CONCLUSIONS

The Australian Cranio Facial Unit with its substantial number of subjects provides a good base for any research on the management of facial clefts. Clinical audits of children treated by the Unit can be used to compare different programs of care over time and to identify trends with in treatment profiles.

This study clearly showed differences and changes in timing of intervention across birth cohort over past five decades. While some of these differences in timing of intervention may be small, others were large and may have a great impact on the future rehabilitation of the children.

The study sample of 255 subjects with facial clefts showed male predominance in comparison to females. The female : male ratio was found to be 1:1.71. The mean age of first intervention for various surgical treatments by birth cohort showed that the mean age has decreased to 3-4 months by the 1990s for repair of cleft lip and variations around the mean has also greatly decreased when compared to earlier birth cohorts. The same was true for repair of cleft palate. This implies that for 1990s birth cohort most of the cleft palate repair were carried out between 6-12 months of age. The most likely age of alveolar bone grafting decreased to 10-12 years.

The most likely age for pharyngoplasty was 4-9 years. Rhinoplasty or nasal tip revision was relatively constant, the mean age of intervention being 9.83 years, but the standard deviation greatly decreased by 1990s birth cohort. The mean age of bilateral myringotomy was also seen to be decreased to 1.80 years in 1990s birth cohort. Osteotomies were seen to be done at mean age of 14 years. Orthodontics were carried out at a mean age of 7.88 years and the deviation around the mean has largely reduced.

This showed that there has been a continuous decrease in the mean age of all surgical interventions through to the 1990s birth cohort. This also implied that most of the surgical interventions employed in the correction of facial clefts are completed much earlier in life compared to earlier birth cohorts. Decreased variation around the mean showed standardisation of procedure for most of the study subjects.

Along with the decrease in mean age and variation in the various surgical interventions, there has also been a decrease in number of days of hospitalisation on account of surgical interventions for correction of facial clefts. Some 31% of the study sample spent 20 days and another 20% spent 10 days and rest of the frequencies ranged from minimum of 6 days to maximum of 50 days. Initiation of speech therapy had also decreased to a mean age of 1.6 years by the 1990s birth cohort. This showed that with time the cost effectiveness of various surgical

procedures and hospitalisation had increased. Presumably success and quality of treatment rendered had been improved.

A protocol for management of cleft lip and palate practised over last 25 years followed by the Australian Cranio Facial Unit was derived.

Retrospective studies like the present study provide the basis for more detailed prospective trials aimed at improving the outcome of treatment. Single centre studies serve well to generate hypotheses for more rigorous research and testing. Such research, when compared with the outcome of studies at other centers, can help in identification of beneficial practises. This should stimulate the development of improved treatment strategies and techniques across all treatment centers which would benefit our patients.

This project also aimed to open further research opportunities in comparing the outcomes to those of other centres. The continued introduction of new techniques and procedures in treatment without careful comparisons against the best currently available alternatives will not serve our patients well (Tulloch, 1993).
More reliable and complete information on the effectiveness of current procedures is required through studies to compare the outcomes of different programs of care.

CLEFT LIP AND PALATE

TIME START ❑❑ TIME FINISH ❑❑

NAME: . . . . . DATE OF BIRTH ❑ ❑ ❑ ACH NO

. . . . . CFU NO

ADDRESS: . . . . . .

GENERAL INFORMATION

AGE IN YEARS ❑❑ COUNTRY OF BIRTH COUNTRY OF BIRTH MOTHER

SEX ❑ COUNTRY OF BIRTH FATHER

ETHNIC GROUP ❑ LOCATION TYPE

1 = URBAN
2 = PRE-URBAN
3 = RURAL ❑

BIRTH WEIGHT BLOOD GROUP

PERIOD OF GESTATION BLOOD GROUP MOTHER

AGE OF MOTHER DURING PREGNANCY BLOOD GROUP FATHER

FAMILY HISTORY OF CL/CP IF YES, WHO (RELATION)

FAMILY HISTORY OF
ANY OTHER G.DEFECT/ANOMALIES

CLEFT CLASSIFICATION

Class I	–	(CL)	–	Cleft of primary palate (Cleft tip =/- alveol are process)	☐
Class II	–	(CLCP)	–	Cleft of primary and secondary palate (cleft up and palate)	☐
Class III	–	CL (P)	–	Cleft of primary palate with or without a cleft of secondary palate	☐
Class IV	–	CP	–	Cleft of secondary palate (isolated cleft palate)	☐

OTHER SYNDROMES AND ANOMALIES

- Cleido cranial dysplasia ☐
- Facial palsy ☐
- Trigeminal neuralgia ☐
- Epilepsy ☐
- Skin lessions ☐
- OTHERS ☐

SPECIFY . . . . . .

. . . . . .

MEDICAL/SURGICAL/DENTAL INTERVENTIONS

SURGICAL

- Rhinoplasty/Septorhinoplasty ☐
- Bone graft to alveolar fistula ☐
- Pharyngoplasty ☐
- Lip Lemesiuriere ☐
- Repair of Cleft Palate ☐
- Repair of Cleft Lip & Alveolous Repair ☐

MEDICAL

- Dermatitis ☐
- Nasendoscopy ☐
- Speech Therapy ☐
- Chest Infection ☐
- Sinusitis ☐
- Pneumonia ☐

DENTAL

- Orthodontic ☐
- Extraction ☐
- Prosthetic Crown/Bride ☐
- Restorative ☐
- Dental Anomalies ☐

- Le Fort I Osteotomy ☐
- Bilateral Myrigotonies ☐

- OTHERS ☐
  SPECIFY . . . . . .

- Meningitis ☐
- ENT Infections ☐
- Otitis Media & effusion ☐
- Asthma ☐
- OTHERS ☐
- SPECIFY . . . . . .

- Occlusion ☐
- Deviation of nasal septum ☐
- OTHERS ☐
- SPECIFY . . . . . .

NUMBER OF DAYS OF ADMISSION

REMARKS

B: BLOOD GROUPING AND STUDY POPULATION PERCENTAGE

Blood Group	Population Percentage
A +	36.4 %
A -	1.7 %
B +	7.5 %
B -	0 %
AB +	4 %
AB -	1.2 %
O +	45.1 %
O -	4 %

C1: MEAN AGE OF FIRST INTERVENTION : REPAIR OF CLEFT LIP

Birth Cohort	Mean	Standard Deviation
1940-1959	0.31	0.04
1960-1969	2.34	4.45
1970-1979	0.60	1.48
1980-1989	0.61	1.42
1990-1993	0.45	0.09

C2: MEAN AGE OF FIRST INTERVENTION : REPAIR OF CLEFT PALATE

Birth Cohort	Mean	Standard Deviation
1940-1959	2.58	3.81
1960-1969	2.27	3.43
1970-1979	1.75	1.80
1980-1989	1.46	1.53
1990-1993	0.68	0.38

C3: MEAN AGE OF FIRST INTERVENTION : ALVEOLAR BONE GRAFT

Birth Cohort	Mean	Standard Deviation
1940-1959	44.0	0.00
1960-1969	12.0	7.00
1970-1979	10.3	3.93
1980-1989	10.3	1.82

C4: MEAN AGE OF FIRST INTERVENTION : PHARYNGOPLASTY

Birth Cohort	Mean	Standard Deviation
1940-1959	17.2	12.8
1960-1969	14.5	9.02
1970-1979	10.5	4.6
1980-1989	6.38	2.63

C5: MEAN AGE OF FIRST INTERVENTION : NASAL TIP REVISION / RHINOPLASTY

Birth Cohort	Mean	Standard Deviation
1940-1959	21.5	10.5
1960-1969	18.6	4.40
1970-1979	9.55	6.18
1980-1989	9.83	2.16

C6: MEAN AGE OF FIRST INTERVENTION : BILATERAL MYRINGOTOMY

Birth Cohort	Mean	Standard Deviation
1940-1959	16.6	9.58
1960-1969	9.78	8.22
1970-1979	4.67	5.16
1980-1989	2.84	2.17
1990-1993	1.80	1.32

C7: MEAN AGE OF FIRST INTERVENTION : OSTEOTOMY

Birth Cohort	Mean	Standard Deviation
1940-1959	26.9	7.25
1960-1969	21.5	5.88
1970-1979	14.1	1.28
1980-1989	14.0	1.99

C8: MEAN AGE OF FIRST INTERVENTION : ORTHODONTICS

Birth Cohort	Mean	Standard Deviation
1940-1959	19.3	0.0
1960-1969	19.8	8.3
1970-1979	11.9	3.9
1980-1989	7.88	2.8

C9: MEAN AGE OF FIRST INTERVENTION : SPEECH THERAPY

Birth Cohort	Mean	Standard Deviation
1940-1959	3.3	2.5
1960-1969	6.1	4.5
1970-1979	4.1	1.7
1980-1989	4.0	2.0
1990-1993	1.6	2.8

D1: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION: REPAIR OF CLEFT LIP

Age	Population Per-cent
3 -4 months	87.4
1 years	6.0
3 years	1.6
4 years	1.4
8 years	1.1
9 years	0.5
10 years	0.5
12 years	0.5
14 years	0.5
43 years	0.5

D2: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION: REPAIR OF CLEFT PALATE

Age	Population Per-cent
6-12 months	74.6
2 years	14.6
3 years	2.2
4 years	2.8
5 years	1.7
6 years	0.6
7 years	0.4
10 years	0.6
12 years	1.1
14 years	0.6
43 years	0.5

D3: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION : ALVEOLAR BONE GRAFTING

Age	Population Per-cent
4 years	5.0
5 years	8.6
8 years	11.4
9 years	14.3
12 years	32.0
13 years	5.7
14 years	11.5
16 years	5.7
19 years	2.9
44 years	2.9

D4: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION : PHARYNGOPLASTY

Age	Population Per-cent
3 years	2.4
4 years	26.0
5 years	2.4
7 years	21.5
9 years	9.5
10 years	9.6
15 years	7.1
16 years	7.1
17 years	2.4
18 years	2.4
21 years	2.4
24 years	2.4
31 years	2.4
45 years	2.4

D5: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION: RHINOPLASTY \ NASAL TIP REVISION

AGE	POPULATION PER-CENT
4 years	2.2
8 years	4.3
9 years	2.2
12 years	34.8
13 years	6.6
14 years	19.5
15 years	4.3
16 years	10.9
18 years	2.2
20 years	2.2
21 years	2.2
22 years	4.3
32 years	4.3

D6 DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION : BILATERAL MYRINGOTOMY

Age	Population Per-cent
1 year	19.0
2 years	27.4
3 years	21.1
4 years	8.4
5 years	12.6
10 years	1.1
12 years	3.2
13 years	1.1
14 years	1.1
15 years	1.0
16 years	1.0
21 years	1.0
23 years	1.0
24 years	1.0

D7: DATA FOR AGE OF FIRST INTERVENTION FOR STUDY POPULATION : OSTEOTOMIES

Age	Population Per-cent
12 years	1.6
14 years	3.4
15 years	44.0
16 years	22.0
19 years	4.0
20 years	8.0
25 years	4.0
27 years	4.0
32 years	6.0
33 years	3.0

E: DATA FOR NUMBER OF DAYS IN THE HOSPITAL FOR VARIOUS SURGICAL INTERVENTIONS RELATED TO CORRECTION OF FACIAL CLEFTS

Days	Population Per-cent
6 days	2.3
8 days	4.0
10 days	20.0
12 days	5.3
14 days	7.4
16 days	4.0
18 days	6.0
20 days	31
26 days	4.0
30 days	11.0
40 days	4.0
50 days	1.0

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