

## CHAPTER 1:

### INTRODUCTION AND ORIENTATION

The aim of the chapter is to review the current best practice for cleft care and the challenges in providing an optimal standard of care for young children with cleft lip and/or palate in developing countries. The rationale for this study is presented. In addition, an overview of the content of the study and justification for terminology used is presented.

#### 1.1 INTRODUCTION

Craniofacial anomalies (CFA) are the fourth most common birth anomalies that occur in approximately 1 in 700 live births (ACPA, 2007: 5; Peterson-Falzone, Hardin-Jones & Karnell, 2010: 15; WHO, 2002: 10). Cleft lip and/or palate (CL/P) is the most common type of CFA and it affects all races across worldwide. The exact cause of CL/P is not known but it is linked to environmental influences, genetic factors and gene-environment interaction (Peterson-Falzone et al., 2010: 38; Watson, Sell & Grunwell, 2001: 10; Wyszynski, 2002: 283). The CL/P causes a pervasive impact on speech, hearing, appearance and cognition. This has a prolonged and adverse influence on the health and social integration of an individual with CL/P. There are considerable costs of cleft care in terms of health care, emotional disturbance, and social and employment factors impacting individuals with CL/P, their families and society on the whole (WHO, 2002: 2).

The management of CL/P involves a multidisciplinary approach and most developed countries have well established protocols for cleft care. The American Cleft-Palate Craniofacial Association (ACPA, 2007), the Eurocran Speech Project (2000) and the World Health Organization (WHO, 2002), have emphasized the need for a well-coordinated, effective team-based approach and early intervention programmes. However, there are numerous challenges to providing cleft care in resource limited developing countries. It is estimated that 80% of the population of individuals with clefts live in the developing or less developed world and may not receive adequate cleft care (Mars, Sell & Habel, 2008: 1).

The surgical team is at the core of management of CL/P in most countries. However, role of the speech-language therapists and audiologists in providing cleft care is not well established in countries with limited resources. It is well recognised that children with CL/P are at risk of communication delays/disorders (ACPA, 2007: 5; Peterson-Falzone, Trost-Cardamone, Karnell & Hardin-Jones, 2006: 9; Rossetti, 2001:3; Strauss, 2004: 150). Early identification and management of communication difficulties by a speech-language therapist and audiologist has a positive impact on the child's overall development (Billeaud, 2003: 53; Bzoch, 2004: 19; Rossetti, 2001: 267; Scherer, D'Antonio & McGahey, 2008: 30). The number of speech-language therapists and audiologists in developing countries is limited (D'Antonio & Nagarajan, 2003: 308). As a result there is a lack of awareness of their contribution to cleft services. These factors clearly indicate a need to establish the roles of speech-language therapists and audiologists at an early stage in the management of CL/P.

In acknowledgement of the many challenges to cleft care in developing countries, clinicians and researchers are currently focusing their attention towards global strategies for improved cleft care. The WHO (2002: 33) recommended international research collaborations and guidelines to improve clinical practice. However, local needs for the cleft care are likely to vary, thus clinical decision-making in speech-language therapy should be guided by research evidence, as recommended by Reilly (2004: 115).

International collaboration is a prerequisite for research into the etiology, prevention and treatment of cleft lip and palate. A critical mass of clinical researchers including epidemiologists and basic scientists is required to generate comprehensive research evidence (WHO, 2002: 2). In order to facilitate international collaboration it is recommended that a set of guidelines for the provision of clinical services and the maintenance and analysis of minimum clinical records are adopted internationally (WHO, 2001b: xi). Developing countries invariably face economic constraints in complying with these recommendations; however, the guidelines should serve as long-term desirable goals in providing an optimal standard of cleft care.

## **1.2 OVERVIEW OF INTERVENTIONS FOR YOUNG CHILDREN WITH CLEFT LIP AND/OR PALATE IN DEVELOPING AND DEVELOPED COUNTRIES**

Health care service delivery models vary across the world and are dependent on the availability of resources. Developing countries differ markedly from developed countries in various aspects of interventions from prevention, identification of developmental concerns to assessments, service delivery, programme evaluation and the formulation of policy. Moreover, even amongst

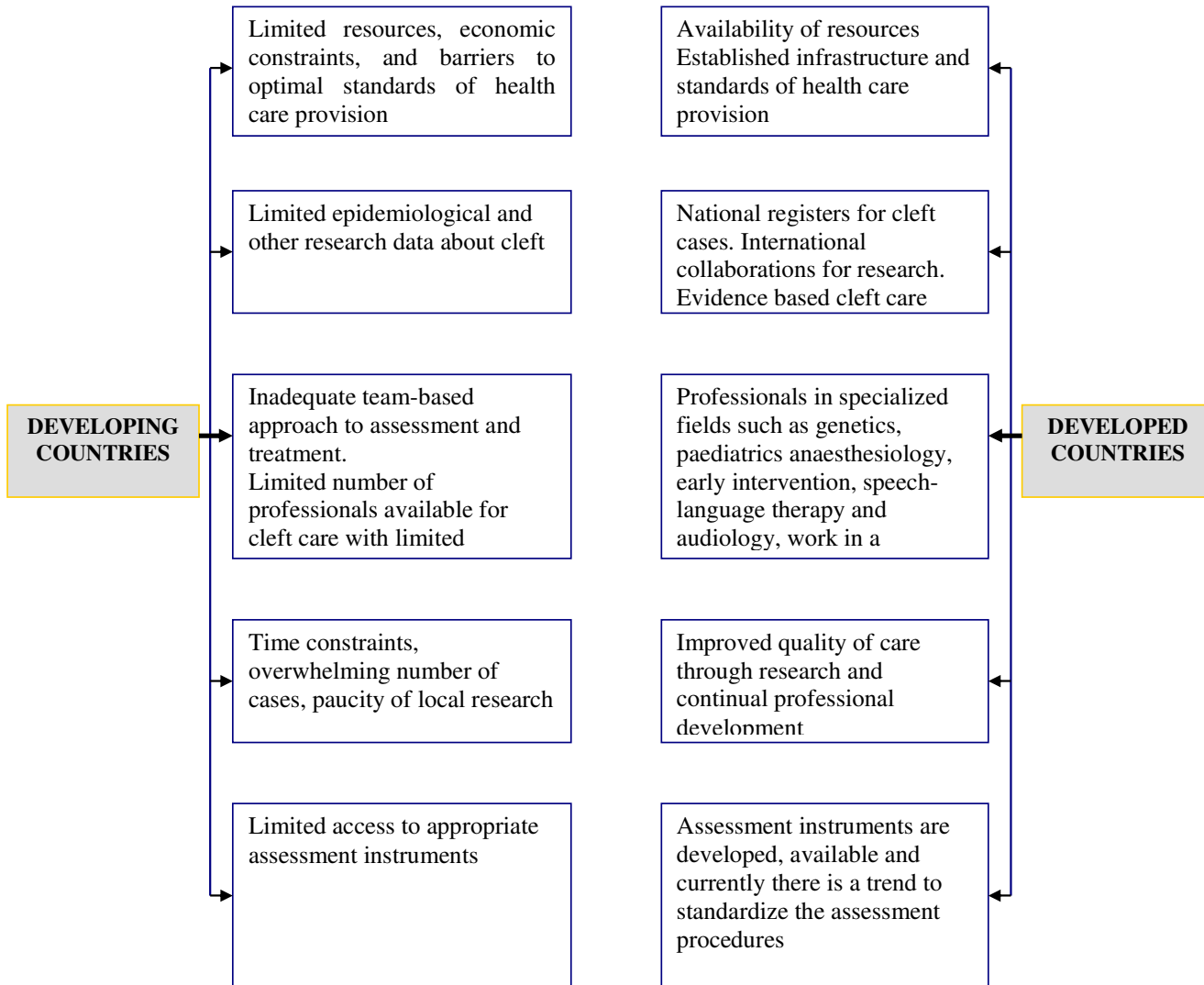
developing countries there are differences in the type of health care delivery (Mars et al., 2008: xi). The common factor in the developing world is the discrepancy between the overwhelming numbers of patients and the limited resources available for their management (Mars et al., 2008: 32). Since many individuals with a cleft have no access to care in developing countries, their cleft may often remain untreated. The priority is undoubtedly to surgically repair the clefts but a long-term interdisciplinary approach is essential to achieve optimum anatomical, physiological and functional results of the cleft repair (ACPA, 2007: 5; Bzoch, 2004: 35).

There are many variations in the treatment protocols used by the various teams all over the world. However, there is consensus that the best outcomes of surgical repair are achieved with a multidisciplinary team-based approach (ACPA, 2007: 5; Bzoch, 2004: 35) and that early repair is associated with positive outcomes for speech development (ACPA, 2007: 14; Peterson-Falzone et al., 2010: 149; Shprintzen & Bardach, 1995: 109; Watson et al., 2001: 162). Surgical repair of the CL/P is effective and available in several developing country settings. Services of visiting surgeons supported by charity foundations such as Interplast, Operation Smile, The Smile Train (Bale, Stoll & Lucas, 2003: 83; Mars et al., 2008: 10) are increasingly available in underdeveloped regions of the world. A major challenge in these contexts is coordinating the timing of surgical intervention and the availability of an inter-disciplinary team. In most developing countries setting up a team for care of young children with cleft may not be possible due to the unavailability or limited number of professionals in specialized fields such as orthodontics, speech-language therapy and audiology.

Children with CL/P exhibit a spectrum of communication problems from delayed speech-language development, to abnormal resonance and speech articulation, and hearing impairment (Kummer, 2008: 299; Peterson-Falzone et al., 2010: 221; Trost-Cardamone, 2004: 463). Evidence from research conducted in developed countries has demonstrated that early communication intervention (ECI) reduces the risk of communication delays and disorders (Guralnick, 1997: 11; Rossetti, 2001: 264; Scherer et al., 2008: 26). However, speech-language therapists and audiologists require a comprehensive communication assessment protocol to assess children with CL/P who may have associated impairment of important functions such as feeding, hearing and developmental delays. The clinical purpose of assessment is to identify the problems, manage them and allow therapists to communicate their findings to other professionals involved in care of children with CL/P (D'Antonio, 2002: 27).

The assessment protocol that is appropriate for the context where it will be utilised needs to take into account the languages and dialects spoken within the child's home and other places of care. Speech measurement procedures across cultures and languages need to be standardised to make meaningful comparisons of clinical outcomes of treatment procedures through multicentre studies to improve the standard of cleft care globally (Henningsson et al., 2008: 1-17). Assessment instruments and procedures for young children with CL/P in developed countries are now being standardised. For example, a universal system for reporting speech outcomes in individuals born with CL/P allows comparisons to be made across centres for either clinical or research use (Henningsson et al., 2008: 1; Lohmander-Agerskov & Olsson, 2004: 64).

Currently, many developing countries do not have access to appropriate speech and hearing services and assessment instruments (D’Antonio, 2002: 1). In the following Figure 1.1, the continuum of cleft care available in developing versus developed countries is depicted from the perspective of speech-language therapists and audiologists.



**FIGURE 1.1 Continuum of cleft care**

Children in sub-Saharan Africa face greater challenges to healthy development than children in any other region of the world (Garcia, Pence & Evans, 2007: 13). In sub-Saharan Africa clinical resources for cleft care are scarce as a consequence of prevailing economic problems and the greater challenge of communicable diseases, particularly HIV/AIDS (WHO, 2002: 38). Mauritius, a small island in the Indian Ocean and forming part of the African continent, is representative of a developing country. Geo-economically it is similar to other developing middle-income countries in sub-Saharan African countries such as Botswana, Namibia, and South Africa. The middle-income economies are defined as ones with a Gross National Income (GNI) per capita of more than \$875 but less than \$10,726. These countries are important because they contribute to the overall economic health of nations as well as to knowledge development and are helping to show the way through political stability and steady economic development (<http://web.worldbank.org>).

The general state of health in Mauritius is good and has been improving steadily over the years. Life expectancy, in the last 30 years has increased from 63 years to 71 years and first year infant mortality has fallen from 64 to 14 deaths for every 1000 live births (Ministry of Health & Quality of Life, 2006). In Mauritius, the government is committed to improve health care during early childhood (0-6 years) through policies that include provision of free primary health care, institution-based facilities that are supported by community-based health workers and free educational services (<http://portal.unesco.org>). Although Mauritius has been doing well in comparison with other developing countries, new measures are required to improve its performance to reach the levels achieved by developed countries. Cleft care in Mauritius is,

however, not yet organised to include interdisciplinary cleft care and a protocol of assessment and treatment is not established.

### **1.3 STATEMENT OF THE PROBLEM**

There are many parts of the world for which little or no information is available on the prevalence of orofacial clefts, in particular parts of Africa, Central Asia, Eastern Europe, India and the Middle East. The lack of information needs to be addressed urgently to establish health care needs of the population (WHO, 2002: 13). In Mauritius, there is a lack of information regarding children with CL/P. The prevalence of children born with CL/P in Mauritius; timing of operations; the investigations and results relevant to CL/P care (feeding, hearing, and speech-language tests) are not documented uniformly. Thus an important first step towards the improvement of cleft services is to channel efforts towards organizing a cleft database and to develop cleft care systems appropriate to the context.

In Mauritius, there is also a need to describe the characteristics of children born with CL/P so that services can be planned and quality care can be provided to these children. There is presently no standardised assessment protocol or guidelines that the speech-language therapists and audiologists follow to conduct assessments of young children with CL/P. Moreover, limited access to assessment instruments undermines the effectiveness of the speech-language therapists' and audiologists' intervention in cleft care. Speech-language therapists and audiologists from countries such as India, Singapore, and sub-Saharan Africa have also voiced the need for realistic assessment tools that have local relevance (Pickering & McAllister, 2000: 94). An important



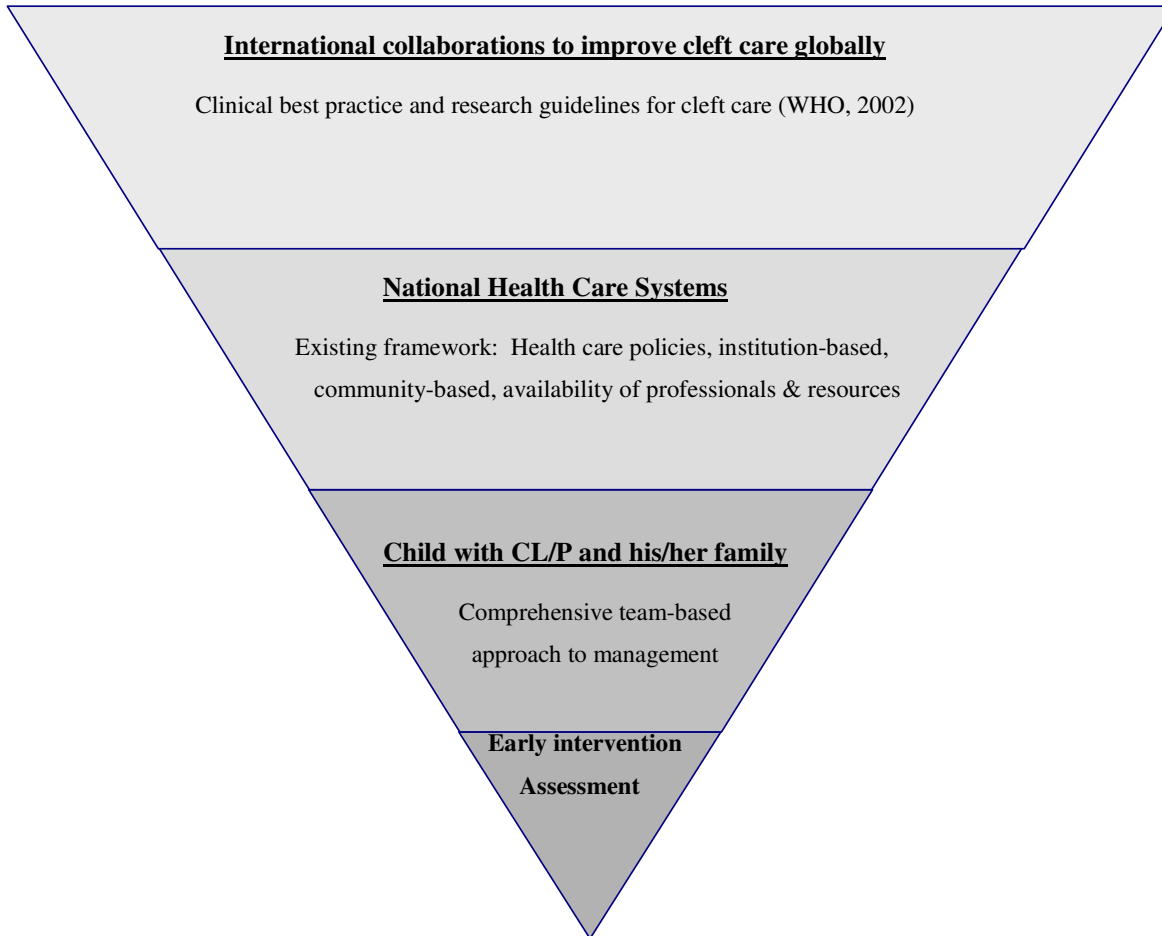
priority in these countries is to develop assessment instruments and treatment materials in local languages based on local experiences and norms.

#### 1.4 RATIONALE FOR THE STUDY

CL/P causes communication disorders and consequently limits the opportunity for education, employment and the development of relationships of the individual (Mars et al., 2008: xi). The WHO (2002) has called for global strategies to reduce the burden of cleft care through standard treatment protocols, improved quality and improved levels of awareness regarding cleft care. However, the access to an optimal standard of cleft care in a developing country is dependent on its resources and health care policies. To improve the health care service delivery to children with CL/P, measure the outcome of surgery and plan efficient service delivery, it is important to have a standard communication assessment protocol. The essential background information concerning individual children should be recorded in a standard and uniform format (CSAG 1998; <http://www.who.int>). Timely interventions by speech-language therapists and audiologists in the period from birth to primary school entry level are crucial for children with CL/P as they are at risk of developing communication delays and disorders (Kuehn & Moller, 2000: 348; Rossetti, 2001: 3).

In a developing country, following the 'bottom-up' approach may focus on the child with CL/P and his/her family as a first step to improving cleft care as illustrated in Figure 1.2. The way to achieve international collaboration is to produce evidence of the needs and the strengths of what

a developing country can offer to improve cleft care. Research is required to follow the route from bottom up.



**FIGURE 1.2 Bottom up approach as a strategy to improve cleft care in a developing country**

A bottom up approach integrates the best external evidence with individual clinical expertise (Threats, 2006: 255). Evidence available from developed countries holds that early communication intervention is beneficial to children with cleft lip and/or palate (Billeaud, 2003:

34; Guralnick, 1997: 11; Hardin-Jones, Chapman & Scherer 2006: 8; Owens, 2001: 458; Rossetti, 2001: 264). In a developing country such as Mauritius, a tangible start to comprehensive cleft care could be early communication intervention. The communication assessment practice should lead to appropriate intervention strategies (Bagnato, Neisworth & Munson, 1997: xiv; Blackman, 1995: 155) and be of immediate value to children with CL/P and their families. However, speech-language therapists and audiologists require an assessment instrument that has local relevance, and documents serial assessments to plan intervention for individual cases with clefts. In addition, access to easily retrievable demographic information and a uniformly recorded assessment database, can help to organise and plan cleft care at a national level (WHO, 2002: 33). The WHO (2001b: ix) has recommended establishing databases of craniofacial anomalies that can be linked for the purpose of international collaborations.

A critical review of clinical practice often helps to generate interesting research questions. (Reilly, Douglas & Oates, 2004: 344). The clinical experience of providing speech-language therapy and audiology services in Mauritius raised questions to be addressed by the research: What is the nature of an appropriate and comprehensive communication assessment protocol for routine clinical use for standard assessment of young children with CL/P in Mauritius? Can a database for individuals with cleft be initiated so that information can be easily retrieved, processed and be used for both clinical services and future research.

The aim of this research study is to develop an appropriate communication assessment protocol for young children with CL/P in Mauritius. A new clinical assessment instrument should be

evaluated prior to routine clinical use to determine its accuracy, acceptability and cost effectiveness. The focus of this study is to apply and evaluate the new communication assessment protocol in terms of its applicability and acceptability for clinical use by speech-language therapists and audiologists. A communication assessment instrument may not find its use in clinical practice if it is not applicable and acceptable to the users in a clinical setting.

## **1.5 THE RESEARCH APPROACH**

In this study an action research approach is adopted as it is particularly suited to identifying problems in clinical practice and helping to develop potential solutions to improve practice (Denzin & Lincoln, 2000: 96). Action research is focused on finding a solution to a local problem in a local setting (Leedy & Ormrod, 2005: 108). The mixed methods design to conduct action research is useful as in a single study; practical questions can be addressed using a combination of data gathering methods, analysis and interpretation approaches (Denzin & Lincoln, 2000: 617-618; Onwuegbuzie & Johnson, 2006: 49). The complementary information derived from quantitative and qualitative data can more effectively address the need for a new communication assessment tool that speech-language therapists and audiologists can use in clinical practice. Furthermore, a participatory action research approach (from a combined quantitative and qualitative approach) is required to utilize existing human resources, to build motivation and awareness and ensure sustainability of the research results. Focus groups, in-depth interviews and participant observations may be used to determine the acceptability of the research study (De Vos et al., 2005: 413).

Speech-language therapy focuses on human communication and social interactions, the orientation in this field is 'social' (Damico & Simmons-Mackie, 2003: 132). The social model of cleft care emphasises the role of the society/community to accept and adapt to a person and the clinician's role to transfer skills, act in a supporting role and to develop extensive family support. (D'Antonio & Nagarajan, 2003: 308; Prathanee, Dechongkit & Manochiopinig, 2006, 503). Contextual, cultural and linguistic aspects are important when conducting speech-language assessments (Carter, Lees, Murira, Gona, Neville & Newton, 2005: 386). Therefore, speech-language therapists in countries with limited resources cannot directly import assessment tools from developed countries. In order to develop/adapt an assessment protocol to improve clinical practice of cleft care in a developing country a critical review of current literature is required. The principles which guide assessment and the recommended assessment procedures for young children with CL/P have been documented (ACPA, 2007). However, these recommendations cannot always be directly applied to clinical practice because there is diversity in the health care systems and the characteristics of the children and families who access these services. The development of a communication assessment instrument requires 'contextualised' and 'authentic' research so that the variables that may act to influence these phenomena may be detailed and analysed (Damico & Simmons-Mackie, 2003: 132). A need therefore exists to conduct empirical action research to develop an assessment instrument that has sound theoretical underpinnings, a computer database, and the instrument is acceptable to the professionals who will utilise the assessment instrument and the electronic data storage tool.

Research is not a priority of health systems in most developing countries. Some general factors inhibiting research in developing countries are:

- Poor health care infrastructure - materials, manpower, political will
- Limited financial resources
- Poor training in research methods, epidemiology and statistics
- Little incentive for busy practitioners who may be overwhelmed due to lack of resources
- Employment systems where only service is rewarded (Horton, 2000: 2231).

In developing countries such as Mauritius, there is a need to focus and prioritize research that will optimize health benefits and prove to be relevant for clinical practice (Reilly, 2004: 121). An instrument based on sound research and developed for a specific community has the potential to be of great sustained value.

## 1.6 TERMINOLOGY

The following frequently used terms within the context of this study are clarified below.

**Craniofacial anomalies:** Craniofacial anomalies (CFA) are a diverse group of congenital deformities in the growth of the head and facial bones. According to WHO (2001a: 10) the term CFA covers a poorly defined group of congenital anomalies that could include any etiologic category (chromosomal, environmental, multi-factorial) as well as any pathogenic mechanism (malformation, deformation, dysplasia) or any clinical category (isolated defect, sequence, syndrome). Craniofacial anomalies are of numerous types; Orofacial clefts that include all types of cleft lip and/or palate are relatively common type of craniofacial anomaly (Mossey, 2005: 31; <http://www.who.int/genomics/anomalies>). Therefore, in this study the term cleft lip

and/or palate, a typical example of CFA has been used and includes isolated forms as well as CL/P associated with a syndrome/sequence.

**Cleft lip and/or palate:** The term includes cases with oral cleft of any type. Some texts use the term CL +/- CP, but the abbreviation CL/P was selected for use throughout the text and includes all types of cleft. A simple and easy to document classification of the CL/P was selected (Bzoch, 2004: 45) as it is based on description of unrepaired clefts of the lip and palate:

- Cleft lip and alveolus (left, right, bilateral)
- Cleft lip and palate (complete, left, right and bilateral)
- Cleft palate (hard, soft and submucous)

In this study distinction between syndromic and isolated orofacial cleft is not made, although the two types have been shown to be epidemiologically distinct (Shprintzen & Bardach, 1995: 7). The communication assessment should be generic; therefore the term CL/P as used in this study includes any type of cleft, isolated CL/P and CL/P associated with a syndrome/sequence.

**Cleft palate speech and language characteristics:** A spectrum of speech-language disorders has been reported among children with CL/P. The cleft type speech characteristics include: hypernasality, inaccurate articulation and frequent glottal stop, nasal emissions during production of fricative sounds. In addition characteristics may include delayed development of language and speech that is accompanied by undesirable facial distortions or mannerisms such as the nasal grimacing (Bzoch, 2004: 404; Hardin-Jones & Jones, 2005: 12; Kuehn & Moller, 2000: 352; Kummer, 2008: 184; Peterson-Falzone et al., 2006: 37; Watson et al., 2001: 235).

**Cleft Palate Interdisciplinary Team:** Refers to a group of multidisciplinary professionals, specialized in the identification, evaluation and management of individuals with cleft who work together to coordinate the patient's care. These specialists include the surgeon, speech-language therapist, orthodontist, ENT specialist, psychologist, paediatrician, social worker, audiologist, clinical geneticist, radiologist, anaesthetist and nutritionist (ACPA, 2007: 7; Kummer, 2008: 301-302; Shprintzen & Bardach, 1995: 14). The use of this term in the study denotes agreement with best practice guidelines as formulated by ACPA.

**Emergent literacy:** Children are in the emergent literacy stage of development from birth through to approximately five years of age. Emergent literacy is defined as the reading and writing behaviours of young children before they become readers and writers in the conventional sense (Justice, 2006: 1-4). These precursors to reading include knowledge about print and books, and have their roots in early home and preschool experiences (Hoff, 2005: 398). This term is used to refer to a specific aspect of communication development of children with CL/P in this study.

**Young children:** The term young children in this body of work will refer to infants, toddlers and preschool children in the age range 0-6 years. Within this group of young children, infants are younger than 12 months, toddlers are between 12 to 36 months and preschool children are in the age range of 3-6 years (<http://en.wikipedia.org/wiki/Infant>). As the early years of life are crucial to fully develop thinking, language, emotional and social skills the focus of this study is young children with CL/P.



**Early Communication Intervention (ECI):** Refers to early intervention services from a communication-based perspective, covering health and all developmental areas that begin at birth and continue to age three years (ASHA, 2008: 1). Early communication intervention includes screening and assessment and is characterised by an emphasis on family involvement and education (Rossetti, 2001: 147; Roth & Worthington, 2005: 136). Early childhood intervention is the term preferred by Shonkoff and Meisels (2000: xii) and it consists of multidisciplinary services provided to children from birth to five years of age to promote health and well being, enhance emerging competencies, minimise developmental delays, remediate existing or emerging disabilities, prevent functional deterioration, and promote adaptive parenting and overall family functioning. In this study, although young children were in the age range of 0-6 years the term ECI was selected to include the key concept of communication intervention for the entire spectrum of early childhood years.

**Communication Assessment:** The term communication assessment as used in this study includes communication skills development, speech, language and hearing. The term 'communication assessment' has an expanded meaning relevant to this study of young children with CL/P as feeding, oral motor functions and developmental aspects are also included. Assessment in early communication intervention is defined as the ongoing procedures used throughout the child's development that include the identification of the child's unique strengths and needs, a family-directed assessment of the concerns, priorities, and resources of the family; the identification of the nature and extent of the early intervention services needed by the child and family; and the identification of supports necessary to enhance the family's capacity to meet the developmental needs of the infant or toddler (ASHA, 2008: 10).

**Communication disorders and delays:** Refers to impairment in the ability to receive, send, process and comprehend concepts or verbal, nonverbal and graphic symbol systems in this study. A communication disorder may be evident in hearing, language and/or speech. Individuals may demonstrate one or any combination of the three aspects of communication disorders. A communication disorder may result in a primary disability or it may be secondary to other disabilities' (ASHA, 1993). It broadly includes all types of speech/language delays, disorders, and disabilities. A speech disorder may be an impairment of the articulation of speech sounds, fluency and or voice and a language disorder is impairment of comprehension and/or use of spoken, written or other symbol systems (ASHA, 1993: 108). The term communication delay in this study refers more specifically to a level of functional communication that is significantly below the expected or typical levels based on a child's age, and refers primarily to speech/language delay.

**Developing countries:** Countries that are defined to be low- or middle-income countries by the World Bank, where living standards are thought to be low relative to high-income countries (World Trade Organisation, 2004) will be referred to as developing countries. Although there is no precise definition, there are thought to be more than 125 countries with populations in excess of 1 million that display these characteristics. There are significant differences in development levels among these countries and even within the same country. The context of this study, Mauritius is classified as a developing country by the International Monetary Fund (United Nations, 2004: 1). In this text the terms developed and developing countries have been used, while recognizing that others such as first/third world countries, minority/majority countries, less

economically/ more economically developed countries may be equally appropriate (Sell, 2007: 13) to the context being studied.

**Speech-language therapists and audiologists:** In the USA, the UK and many other developed countries the practice of speech-language therapy and audiology are two separate specialized fields. However, in developing countries for example in India, South Africa, Brazil and Mauritius, the speech-language therapy and audiology traditionally are combined professions. Therefore, the appellation ‘speech-language therapists and audiologists’ is preferred throughout the text as it denotes the profession as practiced in Mauritius.

## 1.7 ABBREVIATIONS

|            |  |
|------------|--|
| ACPA       | American Craniofacial Cleft-Palate Association   |
| ASHA       | American Speech-Language and Hearing Association |
| CFA        | Craniofacial Anomalies                           |
| CL/P       | Cleft lip and/or palate                          |
| CHRIB      | Clinic for High Risk Babies                      |
| CSAG       | Clinical Scientific Advisory Group               |
| ECI        | Early communication intervention                 |
| GOS.SP.ASS | Great Ormond Street Hospital Speech Assessment   |
| WHO        | World Health Organization                        |

## 1.8 ORGANISATION OF THE STUDY

The research presentation is outlined forthwith.

**TABLE 1.1 Outline and description of the sections comprising this study**

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| <p><b>CHAPTER ONE<br/>INTRODUCTION AND<br/>ORIENTATION</b></p>  | <p>The first chapter provides an orientation to the study, the problem addressed and the rationale for the development of a Communication Assessment Protocol for young children with CL/P, in Mauritius. The terminology selected within the context of this study, a list of abbreviations used and outline of the chapters are also presented.</p>    |
| <p><b>CHAPTER TWO<br/>CLEFT CARE<br/>PRACTICES FOR<br/>YOUNG CHILDREN</b></p>   | <p>Chapter 2 describes the impact of a cleft on the child and his/her family as a backdrop to the current practice of care for young children with CL/P. The existing barriers to providing an optimal standard of cleft care in developing countries are discussed along with strategies to improve cleft care.</p>                                     |
| <p><b>CHAPTER THREE<br/>COMMUNICATION<br/>ASSESSMENT OF<br/>YOUNG CHILDREN<br/>WITH CLEFT LIP<br/>AND / OR PALATE</b></p> | <p>An overview of the current speech, language and hearing assessment practices of young children with CL/P is presented. The need for an appropriate comprehensive communication assessment instrument in developing countries is highlighted and the chapter includes a framework for the development of such a communication assessment protocol.</p> |

**TABLE 1.1 Outline and description of the sections comprising this study (continued)**

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| <p><b>CHAPTER FOUR<br/>METHODOLOGY</b></p>                     | <p>Chapter 4 provides a thorough description of the research methodology employed to develop a communication assessment instrument. It includes the aim of the research, the research design, the research ethics, materials, pilot studies conducted and the training of the speech-language therapists and audiologists to conduct the assessments according to the protocol. Along with the data collection and data analysis the reliability, validity and trustworthiness of the assessment instrument are described in this chapter.</p> |
| <p><b>CHAPTER FIVE<br/>RESULTS AND<br/>DISCUSSION</b></p>      | <p>In chapter 5 the results of the communication assessments of the sample population are analysed, descriptions of the characteristics of the children with CL/P in Mauritius are presented and compared to the literature reviews. The chapter then focuses on the clinical applicability, and acceptability of the protocol by the local speech-language therapists and audiologists.</p>   |
| <p><b>CHAPTER SIX<br/>CONCLUSIONS AND<br/>IMPLICATIONS</b></p> | <p>In this chapter a critical evaluation of the research, conclusions drawn from the research, implications for clinical practice and recommendations for future research are presented. The last chapter closes with a final comment from the researcher.</p>   |
| <p><b>REFERENCES</b></p>                                       | <p>A list of all works to which reference is made in the text is presented.</p>  |
| <p><b>APPENDICES</b></p>                                       | <p>The nine appendices included at the end of the chapter are the ethical clearances, informed consent letters, questionnaires and interview schedules, speech elicitation materials and the Communication Assessment tracking form. The digital video recordings of the children's speech patterns and the focus group discussion are also enclosed.</p>  |

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## 1.9 CONCLUSION

Cleft lip and/or palate is a complex congenital disorder that requires coordinated care by a team of multidisciplinary specialists. Early intervention is crucial to prevent or minimise the negative impacts of this congenital anomaly which may be pervasive. The recommendations for best practice in cleft care are the application of multidisciplinary team-based approach and early intervention to improve the outcomes of treatment of young children with CL/P. However, health service provision and cleft care services vary considerably from developing to developed countries. The concept of a team-based approach to cleft care and early intervention that may be taken for granted in developed countries may be non-existent in many developing countries.

In contexts where individuals have poor access to basic health care, the services offered by the small number of speech-language therapists and audiologists for communication disorders have a different meaning. They are required to share their skills with parents, other health care professionals and community workers (Pickering & McAlister, 2000: 96) to provide and sustain speech-language therapy intervention (D' Antonio & Nagarajan, 2003: 308; Prathanee et al., 2006: 501). With the help of available resources and access to an appropriate communication assessment instrument and an electronic database, speech-language therapists and audiologists in developing countries can meet the challenge of effective cleft care.



## 1.10 SUMMARY

The overview of cleft care in developed countries and the limitations associated with the system in developing countries provided the rationale for the research as well as the research questions and aim of this study. Description of the terms and justifications for their selection, abbreviations and the organisation of the contents provided the orientation to this study.

## CHAPTER 2:

# CLEFT CARE PRACTICES FOR YOUNG CHILDREN

The aim of this chapter is to review the impact of a cleft on a young child and his/her family in order to identify the current best practice of care. Furthermore, the role of speech-language therapists and audiologists in cleft care is critically reviewed with implications for developing countries.

### 2.1 INTRODUCTION

Young children with cleft lip and/or palate (CL/P) have been extensively studied from different perspectives. The causes of CL/P are linked to genetic factors, environmental influences and gene-environment interaction (Peterson-Falzone et al., 2010: 38; Watson et al., 2001: 10). In more than 30% of the cases the cleft may be a part of a syndrome. Children with a syndrome often have associated developmental problems and the documentation of a syndrome increases understanding of the cleft condition (Watson et al., 2001: 91).

Infants with CL/P are at risk of feeding problems, hearing difficulties, communication delays and/or disorders and adverse parent-child interactions (ACPA, 2007: 5; Kummer, 2008: 38; Peterson-Falzone et al., 2006: 9; Strauss, 2004: 150; WHO, 2002: 2). Parameters for evaluation and treatment of patients with CL/P and craniofacial anomalies have been provided by national and international bodies, for example the American Cleft Palate Craniofacial Association (ACPA, 2007) the Clinical Scientific Advisory Group (CSAG, 1998) and the Eurocleft group



(Shaw et al., 2001). The early years (0-6) are critical for children with CL/P, when they require primary surgery for repair of the cleft. The challenge faced by professionals involved in cleft care is to cater for the multiple and diverse needs of both the child and the family, in order to optimize the potential for development despite the congenital anomaly (Bzoch, 2004: 19-20).

The majority of developing countries are unable to provide adequate care for young children with CL/P due to various limitations (refer to Figure 1.1). The World Health Organization (WHO) developed guidelines for improving cleft care globally through an international network for consensus building, planning, protocol development and collaborative research (Shaw, 2004: 238). One of the priorities in developing countries is to establish the health care needs for craniofacial anomalies in the population. This can be achieved by a systematic collection of data and establishing a database/national registry for children with CL/P. In developing countries inadequate ascertainment of the cases is due to many births not being registered. Other reasons are the heterogeneity of cleft cases, the failure to use comparable classification system for CL/P, and lack of agreed criteria for data collection (Shaw, 2004: 241).

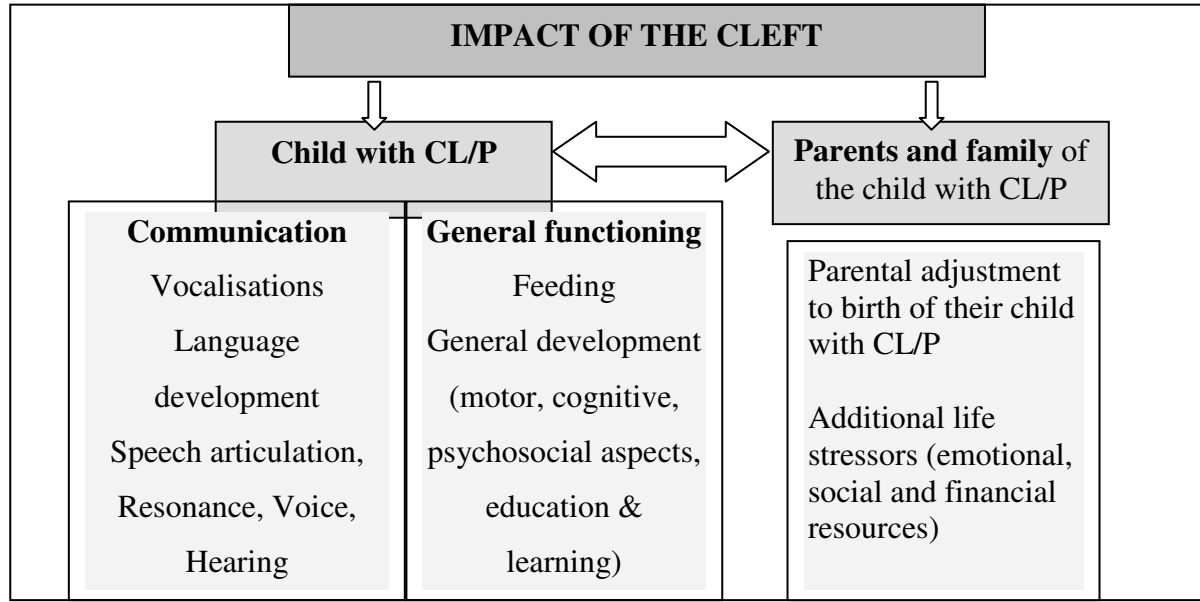
The health and well-being of patients with CL/P is dependent on the clinical expertise of care providers (ACPA, 2007: 5). In many developing countries, scarcity of health care professionals is a major limitation for the application of best practices. It is difficult to plan cleft care services if the information about current care/practice is not known. There is a need to improve cleft care in developing countries to prevent the negative impact of the cleft on a child and his/her family.

Speech-language therapists and audiologists play an important role in improving the standards of care for young children with CL/P in developing countries as they can apply the existing knowledge base of best practice for cleft care from the developed countries to local context. However, are recommended best clinical practices in developed countries, applicable to a developing country? Is there a need to adapt the guidelines to suit the local context without compromising the standards of optimal care? To answer these questions contextual action research is required.

## **2.2 IMPACT OF A CLEFT ON A YOUNG CHILD AND THE FAMILY**

The cleft may adversely affect both the child and the family, but this can be addressed by timely repair, a team-based approach and early communication intervention (ECI). There is no clear relationship between the type and severity of the cleft and its impact on the child and family, as their coping skills and compensatory behaviours are determined by a variety of factors such as temperament of a child, parental coping skills, family's socio-economic status, and the support services available to child and family (Watson et al., 2001: 376).

The overall impact of the cleft on the child and his/her parents, is schematically represented in Figure 2.1 and is discussed.



**FIGURE 2.1 Impact of the cleft**

### **2.2.1 Impact of a cleft on communication development**

Communication skills play a pivotal role in the cognitive, psychosocial, behavioural and social development of a child (Billeaud, 2003: ix). In the USA 6-10% of children younger than three years are reported to have a speech and language development disorder/delay (Billeaud, 2003: x; Rossetti, 2001: 1). In children born with CL/P a higher risk of communication delays/disorders has been reported than children without CL/P (Kuehn & Moller, 2000: 348; Rossetti, 2001: 3). Children with CL/P may also have other risk factors such as the presence of a syndrome, low birth weight (Billeaud, 2003: 55) or HIV/AIDS, to adversely affect their development. The most significant impact of a cleft on communication development is summarised in Table 2.1.

**TABLE 2.1 Impact of a cleft on communication development**

| <b>Communication development<br/>(reported by)</b>  | <b>Impact</b>   | <b>Contributing etiological factors</b>  | <b>Some gaps identified in current<br/>research</b>  |
|---|---|--|--|
| <p><b>Vocalizations</b><br/>(Chapman et al., 2003 : 174;<br/>Hutters, Bau &amp; Brondsted, 2001 :<br/>451-452; Neiman &amp; Savage, 1997:<br/>224; Scherer, 1999: 90)</p>   | <p>Delayed canonical babbling<br/>Reduction in use of consonants<br/>Frequent use of nasal consonants<br/>and glottal stops<br/>Delayed communication<br/>development<br/>Mother-child interactions may be<br/>altered.</p> | <p>Interplay of biological risk factors<br/>(abnormal oropharyngeal structure<br/>and function, intermittent hearing<br/>loss associated with middle ear<br/>effusion) of the child with CL/P and<br/>environmental factors (mother-child<br/>interactions, timing of surgery to<br/>repair cleft)</p> | <p>Insufficient evidence from<br/>research regarding predictive value<br/>of vocalisations to identify infants<br/>at-risk for later speech-language<br/>acquisition in view of the<br/>variability of vocalisations in<br/>infants.</p> |
| <p><b>Language development</b> (Broen et<br/>al., 1998 : 682; Hutters et al., 2001:<br/>456; Kuehn &amp; Moller, 2000: 353;<br/>Morris &amp; Ozanne, 2003: 464 ;<br/>Peterson-Falzone et al., 2010: 241-<br/>242 ; Scherer &amp; D’Antonio, 1995)</p> | <p>Language acquisition is delayed<br/>and/or disordered particularly<br/>among children with syndromes<br/>and sequences</p>   | <p>Hearing and surgical history,<br/>neuro-linguistic deficits, low birth<br/>weight, adverse early<br/>communication interactions,<br/>psychosocial issues and<br/>socio-environmental factors</p>  | <p>Research needed from developing<br/>countries (where there are<br/>inadequate cleft services and lack<br/>of early interventions) on speech-<br/>language development in children<br/>with CL/P.</p>                                  |

**TABLE 2.1 Impact of a cleft on communication development (continued)**

| <b>Communication development<br/>(reported by)</b>  | <b>Impact</b>  | <b>Contributing etiological factors</b>  | <b>Some of the gaps identified in<br/>current research</b>  |
|---|--|--|---|
| <p><b>Articulation</b><br/>(Hardin-Jones &amp; Jones, 2005: 12; Kummer 2008 : 182-191; Persson, Lohmander-Agerskov &amp; Elander, 2006 : 295; Peterson-Falzone et al., 2010: 222-237; Sell, Harding &amp; Grunwell, 1999 : 17-33; Trost-Cardamone, 2004: 463-468)</p> | <p>Weak pressure consonants, nasal emissions accompanying pressure consonants, phonetic errors increase with phonetic complexity, nasal grimace, flare, or facial grimace. Compensatory articulation &amp; obligatory errors</p> | <p>Myriad of structural and learning factors, hearing loss, dental and occlusion problems, phonologic developmental problem, developmental factors (syndromes) and strategies employed to compensate for the cleft</p> | <p>Research needed to examine relationship between articulation, and learning/cognition factors of children with CL/P.<br/>A need to standardize descriptions of articulation, phonologic features and patterns of errors in local languages that have cross-linguistic relevance</p> |
| <p><b>Resonance</b><br/>(Kummer, 2008: 178-182; Trost-Cardamone, 2004: 463-468; Whitehill, 2002: 55)</p>  | <p>Hypernasality: more severe for high versus low vowels and for voiced consonants<br/>Hyponasality: reduced or absent nasal resonance with nasal consonants /m/, /n/, /ŋ/<br/>Mixed resonance<br/>Cul-de-sac nasality</p>       | <p>Velopharyngeal dysfunction, large nasopharyngeal space.<br/>Hypertrophied tonsils or adenoids nasal septum deviation, pharyngoplasty.<br/>Persistent learned habits from the pre-surgery phase</p>                  | <p>Need for refined measures of reliable auditory perceptual resonance analysis and standardisation in local language for use of consistent rating scale of resonance (nasalness varies across languages and dialects)</p>  |

**TABLE 2.1 Impact of a cleft on communication development (continued)**

| <b>Communication development<br/>(reported by)</b>   | <b>Impact</b>   | <b>Contributing etiological factors</b>   | <b>Some of the gaps identified in<br/>current research</b>  |
|--|---|---|---|
| <p><b>Voice</b><br/>(Kummer, 2008 : 190-191;<br/>Peterson-Falzone et al., 2010:<br/>240-241)</p>   | <p>Hoarse voice, unusual<br/>breathiness, and reduced loudness</p>  | <p>Inadequate vocal tract variations to<br/>regulate air pressure for voicing<br/>Compensatory strategy (soft voice,<br/>hyperfunction of vocal cords)</p>  | <p>Need for further research to<br/>conduct differential diagnosis of<br/>voice quality of young children<br/>with CL/P</p>   |
| <p><b>Hearing</b><br/>(Broen et al., 1996 : 132;<br/>Paliobei, Psifidis, &amp;<br/>Angnostopoulos, 2005 : 1373;<br/>Peterson-Falzone et al., 2010:<br/>207; Schonweiler et al., 1999:<br/>277; Shprintzen &amp; Bardach,<br/>1995 : 164 ; Watson et al.,<br/>2001 : 91).</p> | <p>Risk of otitis media and<br/>associated conductive hearing<br/>loss<br/><br/>Sensorineural or mixed hearing<br/>loss</p> | <p>Poor Eustachian tube function<br/><br/>Abnormality of the velopharyngeal<br/>muscles and poor drainage of the<br/>middle ear<br/><br/>Presence of syndrome/sequence<br/><br/>Genetic factors</p> | <p>Research evidence required for<br/>impact of hearing loss on speech-<br/>language development of young<br/>children with CL/P.<br/><br/>The impact of cleft on auditory<br/>processing behaviour of children<br/>with CL/P</p> |

The different areas of communication development that the cleft impacts include: vocalizations, language development, articulation, resonance, voice and hearing.

- **Vocalizations**

The impact of a cleft on an infant's prelinguistic behaviour is important as the infant moves progressively towards speech through the emergence and development of oral motor control, vocalizations, social interactions and language use. Chapman et al. (2003: 175) compared 15 children with CL/P to 15 children without clefts, to determine the relationship between early and later speech development. The findings of this comparative and longitudinal study indicated that children with post cleft repair surgery continued to exhibit speech delays at age 21 months. Thus, children with cleft palate may have fewer 'practiced' forms to call upon for word production. Also, they may receive less reinforcement from parents for communicative attempts as there are fewer instances of canonical babbling for parents to respond to. There is a possibility that parental expectations regarding what sounds the child can or cannot produce prior to cleft surgery may influence patterns of parent-child interactions (Chapman et al., 2003: 192).

The patterns that are evident in children's pre-speech vocalisations persist and are also evident in their early speech (Watson et al., 2001: 195). The prelinguistic consonant inventory of toddlers may impact on the presence or severity of future speech problems (Oller, Eilers, Neil, & Schwartz, 1999: 238; Peterson-Falzone et al., 2010: 235). Hence assessment of early vocalizations is crucial to ECI planning, as specific targets can be set to facilitate early development and include parents in the intervention process.

- **Language development**

Language development is influenced by inherent biological and cognitive abilities of the child and interactions with caregivers (Billeaud, 2003: 33; Guralnick, 1997: 7; Popich, Louw & Eloff, 2007: 65; Rossetti, 2001: 2). Congenital disorders occurring ‘in-utero’ (such as craniofacial disorders) have a more pervasive effect on the learning of speech and language than do similar disorders acquired later in life (Bzoch, 2004: 23).

Expressive language delays have been consistently reported in the majority of children born with CL/P (Kuehn & Moller, 2000: 354). In addition children with clefts may have language delays with the same frequency as children without clefts (Golding-Kushner, 2001: 42). This is understandable as there are multiple risk-factors (established risk factor due to the cleft, biological and environmental risk factors) during the critical period for language acquisition that negatively impact communication development in young children with CL/P (Kritzinger, Louw & Hugo, 1996: 77). A higher prevalence of language delays and speech disorders are reported among children (without cleft) from socio-economically deprived environments compared to their peers (Billeaud, 2003: 45; Guralnick, 1997: 6; Golding-Kushner, 2001: 43; Pamplona et al., 2004: 81). In developing countries children with cleft may be living in poverty and adverse health care contexts, such as inadequate maternal-child care and cleft care services, therefore they have additional risk factors for delayed language development. Research into the interaction between clefting and language development is limited and complex because of the heterogeneity in severity of original deformity and variables such as medical, familial, social and educational factors (Peterson-Falzone et al., 2010: 241). Thus, research evidence for impact of cleft on language development is difficult to find.



In normal speech production the processes of articulation, resonance and phonation are intimately related (Watson et al., 2001: 72) and the impact of the cleft on these are described below.

- **Articulation**

Children with CL/P demonstrate a remarkable variability in articulation performances due to the heterogeneity of cleft types, age groups under consideration and surgical management protocol. However, as a group they often demonstrate poor articulation skills as compared to their peers without clefts (Hardin-Jones & Jones, 2005: 9; Kuehn & Moller, 2000: 352; Peterson-Falzone, et al., 2010: 222). The articulation errors due to structural abnormalities (such as dental anomalies due to the cleft) are termed ‘obligatory errors’ and distinguished from ‘compensatory errors’ made due to a modification in the placement of the tongue and lip movements to compensate for the structural abnormalities (Peterson-Falzone et al., 2010: 224). The place of production of articulation of oral sounds may be shifted to the pharyngeal, laryngeal, and velar loci as a learned behaviour to compensate for the cleft palate (Watson et al., 2001: 198). However, these learnt articulation errors may persist after management of the structural abnormality (Peterson-Falzone et al., 2006: 83). In addition young children with CL/P may have developmental articulation errors (Hutters et al., 2001: 465; Morris & Ozanne, 2003: 464).

- **Resonance**

Despite cleft palate repair surgery, hypernasality may persist in young children with CL/P due to velopharyngeal insufficiency, inadequacy or dysfunction, palatal fistula and/or mislearning (Kummer, 2008: 192). Hypernasality is perceived in connected speech and in vowel production

and its negative impact is increased with the rapid rate of speech and muscular fatigue. Other resonance disorders due to the cleft that may impact on speech intelligibility of children are hyponasality and mixed nasality. Hyponasality may be due to nasopharyngeal obstruction (hypertrophied adenoids), maxillary retrusion as a phenotypical feature of a syndrome (for example, Crouzon syndrome, Apert's syndrome), or due to complications of the surgery conducted to reduce the velopharyngeal gap. Mixed resonance may occur when both velopharyngeal dysfunction and nasal blockage are present (Kummer, 2008: 182). Moreover, nasal emissions, turbulence and grimace often occur simultaneously and have similar etiology to resonance problems and articulation errors. These overlapping problems may be due to learned behaviours 'habituated nasal and facial grimacing' or due to velopharyngeal dysfunction (Bzoch, 2004: 406) and are visually distracting in communication.

Resonance varies across languages and dialects and this may have an impact on how far the child's resonance deviates from 'normal' nasal resonance. Knowledge of the language and/or dialect is essential to determine typical or deviant resonance. Speech-language therapists and audiologists are faced with a challenging task to delineate the contributing etiological factors of resonance, articulation and voice that often co-occur in children with CL/P. Determining the etiological factors contributes to making appropriate management decision (for example physical treatment or behavioural modification).

- **Voice**

In a study by Hocevar-Boltezar, Jarc and Kozelj (2006: 27), voice abnormalities such as hoarseness, unusual habitual pitch, breathiness and reduced loudness were reported among

12.5% of the children with CL/P. Children with CL/P and velopharyngeal inadequacy are at risk of hoarseness due to vocal hyperfunction. The interaction of velopharyngeal inadequacy and the laryngeal compensatory behaviour may result in gottalisation of the stop consonants and it has been hypothesized that these may be the cause of voice disorders in children with CL/P (Bzoch, 2004: 409; Peterson-Falzone et al., 2010: 240). The impact of a cleft on voice quality should be assessed (hoarseness may indicate underlying velopharyngeal insufficiency) and appropriate intervention provided (Kummer, 2008: 190; Peterson-Falzone et al., 2006: 36).

- **Hearing**

Research spanning over years has provided evidence that children with CL/P are at risk of recurrent otitis media and associated conductive hearing loss due to poor Eustachian tube function (Broen et al., 1996: 132). Ear disease, congenital malformations of the auditory system, and congenital sensorineural or mixed hearing loss are frequently seen in children with multi-anomaly disorders such as Pierre Robin sequence, Treacher Collins syndrome, Stickler syndrome, Velo-cardio-facial syndrome and Crouzon syndrome (Shprintzen & Bardach, 1995: 19). The extent and type of hearing loss in children with CL/P is reported to vary, depending on the age, pre and post palatoplasty status and other genetic defects of the sample population (Peterson-Falzone et al., 2010: 209).

In a study by Schonweiler et al. (1999), of 417 of children with cleft palate, it was reported that 80% of the children had hearing problems predominantly of a fluctuating conductive nature caused by otitis media with effusion. In a recent study of 40 infants with CL/P, a moderate hearing loss was found in 35% of the infants, indicating that these infants may be at risk of

speech-language delays/disorders (Andrews et al., 2004: 10-17). The incidence of hearing loss in children with CL/P, in developed countries where audiology and ENT services are available is approximately 58% (Kemker & Antonelli, 2004: 357; Merrick, Kunjur, Watts & Marcus, 2007: 532). In countries where preventive measures such as early myringotomies and placement of ventilation tubes or early intervention for hearing loss among children with CL/P, are not practiced, the incidence is likely to be even higher.

Middle ear disease and hearing loss constitute a major risk for communication delays and disorders in children with CL/P, as even a mild fluctuating hearing loss may have a negative impact upon speech and language (Schonweiler et al., 1999: 215). Early identification and management to protect otologic and audiologic function is critical to the normal development of cognition, language and speech (Mars et al., 2008: 212; Merrick et al., 2007: 532).

An important aspect of hearing is functional listening. Hugo, Louw, Kritzinger and Smit (2000: 47-53), identified the need for a simple and quick tool for the evaluation of listening behaviour that could be easily applied to the high risk population between birth and three years of age and guide early intervention efforts. Yet, auditory processing disorders in young children with CL/P have not been the focus of research to date.

Knowledge of the impact of clefting on communication, language and speech production in young children with CL/P is important as speech-language therapists and audiologists can conduct appropriate assessments and share the results with parents and other professionals involved in cleft care to make appropriate management decisions.

## 2.2.2 Impact of a cleft on general development

The cleft condition can also affect other functions of the developing child with CL/P namely feeding and general development such as motor development and psychosocial aspects. These associated problems may also negatively impact on communication development of the child and are viewed as stressors to the child's family (Watson et al., 2001: 192).

- **Feeding**

The cleft palate may have a negative impact on the neonate's feeding skills (Reid, Kilpatrick & Reilly, 2006: 702) which in turn may have an adverse effect on the adequacy of nutrition, cause stress to parents, adversely affecting mother-child interactions and may also potentially affect oral-motor and oro-sensory development (Kummer, 2008: 121; Reid et al., 2006: 702; Shprintzen & Bardach, 1995: 63). Feeding difficulties of neonates with cleft palate may include poor oral suction, poor intake with lengthy feeding times, nasal regurgitation, and choking, gagging and excessive air intake (Kummer, 2008: 127; Reid et al., 2006: 702; Shprintzen & Bardach, 1995: 65). The primary concern of most parents when an infant is born with CL/P is feeding due to its importance for survival and growth. During the first few months of any infant's life, parent and other caregivers find that most interactions involve feeding and communication (Arvedson & Brodsky, 2002: 527; Young, O'Riordan, Goldstein & Robin, 2001: 55).

Chatoor et al. (1997: 80) and Reid et al. (2006: 702) reported that the cleft palate may not be the main or only cause of the feeding difficulties. Other factors such as the presence of a sequence,

syndrome, prematurity, low birth weight, cardiac or pulmonary disease, functional or structural abnormalities of the oro-pharynx or gastrointestinal tract may contribute to the feeding difficulties. Anomalies such as micrognathia, macroglossia and neuromuscular coordination problems may cause swallowing difficulties which are exacerbated by the cleft. Several secondary problems may occur due to feeding difficulties such as poor weight gain, lengthy feeding times, and stressful feeding interactions between the infant and caretaker (Kummer, 2008: 128; Shprintzen & Bardach, 1995: 63-74).

The anatomical structures for feeding, swallowing and speech are the same although the neurophysiological function differs (Peterson-Falzone et al., 2006: 11). Speech-language therapists are trained to and can offer guidance to parents regarding feeding problems in their children with CL/P. Information concerning feeding is of the highest priority to parents of neonates with CL/P (Young et al., 2001: 57). They may require support, information and intervention concerning feeding from professionals such as speech-language therapists and nursing personnel.

- **General development**

Infants with clefts may exhibit other risk factors such as a syndrome, sequence, congenital malformations, associated anomalies, and environmental risk factors that interact in a synergistic fashion and place them at double risk for developmental delays (Neiman & Savage, 1997: 218; Peterson-Falzone et al., 2010: 376; Shprintzen & Bardach, 1995: 182). Literature has been inconclusive regarding the outcome of general development of infants with CL/P because clefts present as a heterogeneous group of impairments.

In a comparative study of infants with CL/P matched to infants without clefts, Neiman and Savage (1997: 223-224) reported slower developmental performance among infants with CL/P in motor, self-help, cognitive and expressive language domains. Kritizinger, Louw and Hugo (1996: 83) also provided data to support their view that infants and toddlers with CL/P are at risk for developmental delays. Therefore, screening of general development is required to identify children with CL/P and guide appropriate interventions.

In the preschool years (3-6 years), the cleft condition may have an impact on the child's self-image and socialization (Peterson-Falzone et al., 2010: 378). Psychosocial aspects include a range of aspects such as psychological functioning, personality and adjustment, self concept, body image and satisfaction with appearance, social functioning, development and learning (Broder, 1997: 402; Peterson-Falzone et al., 2006: 14). The heterogeneity of aspects that the various studies focus on and the variety of methodologies used (observations, questionnaires, interviews) raises the question whether young children with CL/P have a higher prevalence of psychosocial problems. This question has not been answered conclusively to date.

Hunt, Burden, Hepper and Johnston (2005: 274) conducted a systematic review of published scientific research on the psychosocial impact of CL/P among children and adults. Their study analyzed 64 articles and the conclusion was that overall adjustment and functioning in children with CL/P appears to be reasonably good. The ability to communicate plays a crucial role in the development of appropriate psychosocial skills and behaviours (Hauer, Shriberg, Kwiatkowski & Allen, 2005: 636). Therefore, poor speech intelligibility in children with CL/P may have a negative impact on the speaker's self concept and may affect social development. Limited

research findings on psychosocial aspects are available especially from contexts where cleft care may be of inadequate standard. In developing countries where plastic surgery is not available/accessible to all children with CL/P, or surgery is performed late, the facial appearance due to an unrepaired CL/P may cause irretrievable damage to the psychosocial well being of the individual.

Children with CL/P are also at risk of learning disability, low school achievement, reading difficulties and grade retention due to cognitive deficiencies that are reported to be secondary to language disorders, speech problems, and psychosocial underachievement (Broder, Richman & Matheson, 1998: 129; Richman & Ryan, 2003: 154). In a multicentre study conducted by Broder et al. (1998) to determine the prevalence of learning disability amongst children with CL/P it was found that 46% of the participants with clefts had learning disability, 47% made poor educational progress and 27% had repeated a grade in school. Although the educational difficulties may be attributed to the presence of a syndrome and cognitive deficiencies, research indicates that even children with nonsyndromic CL/P are more likely to have learning problems when compared to children without clefts. Team members are required to be sensitive to the impact of a cleft on the education of young children with CL/P and help to develop effective psychosocial and educational strategies to enhance and support the learning experiences of these children (ACPA, 2007: 22).

Thus, a cleft may have an adverse effect on a child's health and social integration, due to the multiplicity of etiologic factors related to the cleft that impact on communication, appearance, and general developmental functions.



### **2.2.3 Impact of a cleft on parents and families**

The birth of an infant with a cleft is a shocking and traumatic experience for parents. They may experience feelings of sadness, guilt, anger and fear for their child's appearance (Peterson-Falzone et al., 2010: 371; Strauss, 2001: 227; Watson et al., 2001: 118). Parents are often aware of the cleft condition and/or presence of a syndrome in their to-be born child as prenatal diagnosis with imaging techniques is possible in many developed countries. However, the subject of concern is how this information is communicated to parents. An experienced professional who cares should communicate the prenatal diagnosis, and then the family has the opportunity to work through much of their distress before the infant is born (Ter Poorten & Louw, 2002; 56-67; Watson et al., 2001: 118). Strauss (2001: 230) suggests that health care professionals portray children with congenital conditions in positive terms to encourage the family and community to become more accepting of these children.

The infant's feeding difficulties and maintaining adequate weight gain may have a further negative impact on parental coping strategies (Young et al., 2001: 57). The cleft may also impact mother-infant attachment and interactions (Speltz et al., 1993: 487). However this is a controversial topic as recent studies (Baker, Owens, Stern & Willmot 2009: 234; Maris, Endriga, Speltz, Jones, & DeKlyen, 2000: 262) have shown that enhanced personal and social resources helped parents cope with the stress and consequently resulted in positive psychosocial outcomes for young children with CL/P.

Parents of infants with CL/P show elevated levels of stress during infancy and toddlerhood. Early parenting stress was associated with higher levels of adjustment problems when children reached toddlerhood (Pope, Tillman & Snyder, 2005: 558). Parents may also have to balance their time among siblings of the child with CL/P and the siblings need to understand that the young child with CL/P may require longer time to feed, and have several medical visits (Kummer, 2008: 281). Parental stress may relate to accessing professionals and community services, securing adequate financial resources and coping with the stress of sending a child for surgery (Collett & Speltz, 2006: 264). The post surgical period stressors that parents of children with CL/P experience are related to aspects such as a feeding plan, arm splints and pain management. As the child grows, the parents may have other concerns such as the child's fluctuating hearing loss, speech problems, and scholastic and behaviour problems. Clefts may be associated with syndromes such as the Velocardiofacial syndrome (VCFS) that is known to be associated with learning difficulties and later psychological disorders (Kummer, 2008: 103; Peterson-Falzone et al., 2010: 72; Watson, 2001: 263).

In addition to the above factors, reactions of extended family members and friends will have an effect on the child and his/her family and the support that parents may receive in caring for the child with CL/P (Peterson-Falzone et al., 2006: 10). The adverse effects on family functioning can be minimised through family-focused and community-based support services (Rossetti, 2001: 268).

The WHO (2002: 28) recognizes the burden of care to the families of children with CL/P and to society as a whole. Elucidating the pervasive and negative impact of the cleft on

communication, appearance, and general development functions of the child and its impact on parents, provides the underpinnings for a description of the current recommendations for best clinical practice.

### **2.3 RECOMMENDED BEST PRACTICE FOR THE CARE OF YOUNG CHILDREN WITH CLEFT LIP AND/OR PALATE**

Recommendations for the optimal care of young children with CL/P have been made by various groups across the world such as the Clinical Standards Advisory Group (CSAG, 1998) in the UK, the Eurocran group (2000), the World Health Organisation (2001) and the American Cleft Palate and Craniofacial Association (ACPA, 2007). As illustrated in Figure 2.2 recommendations for care a young child with CL/P and his/her family include a team-based approach, early communication intervention and treatment outcome measures. The WHO (2001 b: ix) has recommended the establishment of local databases of children with clefts in order to organise and plan cleft care services and facilitate inter-centre and international collaborations to improve cleft care globally.

|  |
|--|
| <b>RECOMMENDATIONS FOR A CHILD WITH CL/P AND THE FAMILY</b>  |
| <b>Team-based approach to assessments and treatment</b>  |
| <b>Early communication intervention and parental participation</b>   |
| <b>Treatment outcome measures</b>  |
| <b>(ACPA, 2007)</b>  |
| <b>National registry and databases on individuals with craniofacial anomalies linked to global registry on craniofacial anomalies (WHO, 2001b: ix)</b> |

**FIGURE 2.2 Recommended best practice for the care of young children with CL/P**

The recommendations depicted in Figure 2.2 are discussed in the following sections.

### **2.3.1 Team approach**

Children with CL/P require surgical repair of the congenital defect, and cleft care involves a variety of specialists representing many disciplines, working in an interdisciplinary and coordinated team. A team approach to care for young children with CL/P increases the awareness and treatment of the full spectrum of health problems associated with these anomalies. Moreover, comprehensive assessments conducted by experienced professionals in cleft care, and long term follow-up results in the best outcomes of cleft care (ACPA, 2007: 7; Kummer, 2008: 299). In a team approach, professionals specialized in cleft care, assess and provide coordinated treatment to children with CL/P and care for their families, and engage in inter-professional communication within the team (ACPA, 2007: 8; CSAG, 1998: 28). A team approach allows for a comprehensive assessment, with fewer visits which makes the care of patients easier for providers, yet more effective for the patients (ACPA, 2007: 5; CSAG Report, 1998: 21; Nackashi, Dedlow & Dixon Wood, 2004: 269-279; Shaw et al., 2001: 9; Shprintzen & Bardach, 1995: 12-15; WHO, 2002: 143; Wyszynski, 2002: 293). An important concept in the team approach is the inclusion of parents as full team members in the assessment and management process of young children with CL/P.

In order to provide quality care, the team members need to have special training in cleft care (Kummer, 2008: 305). Furthermore, the number of patients referred to the team should be sufficient (at least 40-50 new cases annually) to sustain the experience and specialist skills of all

team members (WHO, 2002: 142), most importantly the surgeon. The Clinical Standards Advisory Group (1998: 98) in the UK recommended that cleft services be centralized to allow fewer centres to function as high volume, high quality ‘hubs’ where experienced team members would provide assessment and diagnostic services while outreach clinics (spokes) conduct follow-up and therapy. Although high volume centres may not guarantee a good outcome, they do provide the means whereby outcomes can be monitored and assessed (Watson et al., 2001: 59). Such a model of cleft care service delivery could be considered to maximise the use of limited professional resources.

The main barrier to following a team-based approach for treatment of individuals with CL/P, in developing countries, is that the professionals who represent the cleft care team are either unavailable or limited in number. For example, in developing countries the availability of surgeons experienced in cleft surgery is a major concern and limited hospital facilities and services may exist (Wyszynski, 2002: 424; Yeow et al., 2002: 18). International teams sponsored by charity organisations (such as Interplast, Rotoplast, Operations Smile, The Smile Train, Transforming Faces) provide surgical interventions in countries where the facilities for repair of the cleft are not available (Mars et al., 2008: 10). However, the dental and orthodontic services for children with CL/P require long term follow-up. These services are not readily accessible or affordable to children with CL/P in developing countries. Speech-language therapy services are often absent or extremely limited in developing countries (D’Antonio & Nagarajan, 2003: 307; Sell, 2007: 14). Training of health care professionals regarding aspects of speech-language therapy and improved level of awareness of cleft care were some of the strategies applied during the Smile Train initiated Pan African Congresses on Cleft Lip and Palate

(PACCLIP) held in Nigeria (2006, 2007) and Ethiopia (2009), to improve cleft care in Africa (<http://www.adventisthealthinternational.org>).

### **2.3.2 Early communication intervention and parental participation**

Early Communication Intervention (ECI) is crucial for young children with CL/P as both biological and environmental factors place these children at risk of communication delays/disorders. Biological risks include the cleft, its associated anomalies and hearing impairment. In addition to these biological risk factors, environmental risk factors affect communication development namely psychosocial issues, early mother-child communication interactions, and family stressors such as financial aspects and social supports (Guralnick, 2005: 14; Kritzinger et al., 1996: 77; Kuehn & Moller, 2000: 354; Savage, Neiman & Reuter, 1994: 222). ECI is reported to promote child health and well being, enhance emerging competencies, minimize developmental delays, remediate existing or emerging disabilities, prevent functional deterioration and promote adaptive parenting and overall family functioning (Shonkoff & Meisels, 2000: xvii).

In developed countries, the role of speech-language therapists and audiologists in ECI has been expanded to include prevention of communication delays and disorders (ASHA, 2008: 9). It is easier and more efficient to prevent the development of speech and language problems than to treat them after they have occurred (Golding-Kushner, 2001: 46). Speech-language therapists and audiologists assess feeding, hearing, prelinguistic communication assessments, parent-child interactions and provide interventions from the neonatal stage of a child with CL/P (ACPA,

2007: 6). Early monitoring and preventive measures have the potential to enable the child with a cleft to achieve normal speech development and hearing acuity by five years of age (Blakely & Brockman, 1995: 25).

Health care professionals can provide appropriate and effective clinical treatment for CL/P and need to empower parents of children with CL/P through education and training (Labuschagne & Louw, 2005: 117; Watson et al., 2001: 379). The most important aspect in the care of young children with CL/P is the partnerships between parents and health care professionals (Nackashi, Dedlow & Dixon Wood, 2004: 274). Young children with CL/P were reported to benefit from early language intervention, such as naturalistic intervention models, enhanced milieu teaching and focused stimulation implemented by early interventionists, speech-language therapists and parents (Pamplona & Ysunza, 2000: 231; Scherer & Kaiser, 2007: 359).

As mentioned previously, the recommended best practice for cleft care is evaluation and treatment by a transdisciplinary team, from the moment of birth in partnership with the parents of the child (ACPA, 2007: 6). However, outcomes of these expensive health care models need to be measured so that scientifically derived findings guide best clinical practice (Reilly, 2004: 113). Moreover, effective ECI requires local speech-language therapists and audiologists with cultural and linguistic competence. But in developing countries only a limited number of speech-language therapists are available who cannot meet the needs of the population. A possible solution to this problem is the development of training programmes for the professions in countries where it has not existed before but this proposition is very costly and requires long term planning (D'Antonio & Nagarajan, 2003: 308). An inspiring example is the Sri Lankan

Cleft Lip and Palate Project that became the catalyst for the development of the profession of speech-language therapy in that country (Mars et al., 2008: 108).

### **2.3.3 Treatment outcome measures**

Highly complex and varied protocols are used to treat individuals with CL/P by different teams. The Eurocleft survey (Shaw et al., 2001: 1) showed that there were 194 different protocols followed for unilateral clefts in Europe alone. Evidence-based practice integrates research findings with clinicians' experiences and patient preferences (Reilly, 2004: 113). If decisions on intervention are to be based on evidence, it is important that treatment outcome measures are in place to judge the effectiveness of the treatment, to compare the results and to improve the quality of cleft care (Watson et al., 2001: 386).

Currently in cleft care, the important measures of treatment outcome are the degree of handicap that persists despite surgical treatment, such as dento-facial development, speech, and psychosocial well being of the child with CL/P (Wyszynski, 2002: 433). From the perspective of clinicians, monitoring and tracking treatment outcomes requires the use of appropriate, consistent and reliable recording of assessments and of interventions (ACPA, 2007: 24; Phillips, 2004: 297).

Speech has been identified as a key outcome measure in children with CL/P (Kuehn & Moller, 2000: 369; Sell, 2005: 116; Lohmander-Agerskov & Olsson, 2004: 68; Persson, Lohmander-Agerskov & Elander, 2006: 307). Speech outcome measures are crucial and need to be



conducted periodically to assess speech at stages of growth and development; for example 0-3 years; preschool age, school age and in adulthood (Kuehn & Moller, 2000: 369). However, the main challenge is to define the speech characteristics to be measured and develop methods that are valid and reliable for how the speech characteristics will be measured and compared (Henningsson et al., 2008: 4; John et al., 2006: 273; Lohmander-Agerskov & Olsson, 2004: 65). Recently the Universal Parameters for Reporting Speech Outcomes (Henningsson et al., 2008:1-17) were developed by a group of speech-language therapists as a tool to measure speech outcome and allow international comparisons for individuals with CL/P. Although this continues to elicit debate (Lohmander-Agerskov, 2008: 452) an important step to standardise speech outcome measures has been taken. In order to guide evidence based practice of cleft care, speech-language therapists require explicit criteria of standardized outcome measures for speech and access to reliable and measurable speech outcomes (John, Sell, Sweeney, Harding-Belle & Williams, 2006: 272; Sell, 2005: 105).

An additional perspective of treatment outcomes that guides best practice is the satisfaction of patients and their families with the treatment provided (ACPA, 2007: 6; Kuehn & Moller, 2000: 370). Quality of life is challenged in individuals with CL/P as a result of impaired function, appearance and social integration. To develop and standardize psychological and quality of life outcome measures, there is an urgent need to either create collaborative groups or improve the networking capabilities of existing groups (Prahl & Prahl-Andersen, 2007: 120; WHO, 2002: 97).

Outcome measures for cleft care that are undertaken on a national/international basis can provide robust evidence to governments for implementing major changes regarding the delivery of cleft care. The development and standardization of valid outcome measures in the treatment of children with CL/P was identified as an important strategy to improve cleft care globally (ACPA, 2007: 6; Bearn et al., 2001: 33; Watson et al., 2001: 391; Wyszynski, 2002: 433). However, a challenge to determining outcome measures for children with CL/P is that interventions are provided at an early stage of life and their consequences are revealed some years later (Wyszynski, 2002: 434). This requires longitudinal, long-term follow-up of the individuals with CL/P and a database can greatly assist in this endeavour. Moreover, as the CL/P may impact many structures and functions, the quantification and weighting of diverse outcomes is required (WHO, 2002: 26). Outcome studies require large data sets that can be used as a basis for outcomes data in clinical trials investigations (Hathorn et al., 2006: 404; Henningson et al., 2008: 15). However, obtaining large data samples may be problematic unless there is international collaboration (WHO, 2001b: 13). Most studies and reviews on CL/P conclude that more research is needed to validate the findings as there is a lack of uniform outcome measurements (Prahl & Prahl-Andersen, 2007: 218). Despite the many issues related to outcome measures it is clear that best practice will only evolve if valid outcome measures that have contextual relevance are in place (Bearn et al., 2001: 42).

#### **2.3.4 National registry and databases for individuals with craniofacial anomalies**

The WHO (2001b: 45) has formulated guidelines to formalize and standardize population based birth-defect registries at national and regional level to contribute to improving cleft care globally.

Access to such statistics and epidemiological information concerning young children with CL/P is crucial to the planning of health care services, prevention, international comparisons and collaboration. The rationale for the registry of individuals with craniofacial anomalies is that it will identify global variability in the prevalence of craniofacial birth defects, estimate the burden of need for public health services, identify priorities and underpin research initiatives that will address primary, secondary and tertiary prevention.

Appropriate documentation and uniform record keeping of individual cases are essential to set up a national congenital anomalies' register using a computer-based data system. However, in developing countries recording of congenital anomalies at a national level may not be possible unless there is a legislation to make such data recording compulsory and there is close collaboration between the public and private health sectors with the National Central Statistics Office (Hammond & Stassen, 1999: 155). A practical way of ascertainment of cases with CL/P would be from the medical/surgical files. Four established networks for registering birth defects are the ECLAMC in Latin America, the EUROCAT, in Europe, the NBDPN in North America, and the ICBBD located in Rome which is the most widespread programme as it includes 34 countries across the 5 continents (WHO, 2001b: 48). These systems could serve as models for the setting up of a national database that could be linked to an international database. Creation of national registers and international databases are of primary importance if developing countries are to strive to provide best practice for children with CL/P.

The evolution of computerized records may facilitate the challenging task of creating databases but the maintenance of a reliable national register system is difficult, time consuming and

expensive (WHO, 2001b: 5). An organised register of individuals with CL/P should allow the required information to be easily retrieved, processed and used for both clinical services and future research. Furthermore, setting up a global registry of cases with CL/P and craniofacial anomalies maximizes opportunities for preparatory work on outcome measures (WHO, 2002: 30).

While developed countries continue with their efforts to improve the standard of care for young children with CL/P, the current reality in developing countries is that more pressing health care priorities such as malnutrition, under nutrition and communicable diseases such as HIV/AIDS overshadow the care of children with CL/P (Mars et al., 2008: 1; WHO, 2002: 36). Furthermore, the financial burden of care and problems of access to professionals in cleft care put treatment beyond the reach of vast numbers of individuals with CL/P in developing countries, (WHO, 2002: 2). However, by addressing the specific barriers to providing cleft care in developing countries and by suggesting strategies based on the guidelines of best practice these challenges may be overcome.

Research from developed countries can inform and provide the theoretical underpinnings for improving cleft care in developing countries. For example Scherer et al. (2008: 25) provided evidence that mothers of children with CL/P could be trained to deliver intervention reliably under the guidance of speech-language therapists. This has implications for developing countries where the number of speech-language therapists is limited. However, there is a need to conduct local empirical research to adapt, and/or develop identification and subsequent management programmes for children with CL/P that have socio-cultural relevance (D'Antonio

& Nagarajan, 2003: 310; Sell, 2007: 17). Contextual research is required to determine the potential of utilizing family centred intervention (Louw et al., 2006: 48).

## 2.4 CONCLUSION

The negative impact of a cleft on a child's oral structures (velopharyngeal closure, dentition), development (feeding, communication skills development, psychosocial development, and education) and auditory system are well known. Professionals from various disciplines have agreed that to meet multiple and complex needs of children with CL/P and their families the best practice for cleft care requires:

- An interdisciplinary, team-based approach to assessments and treatment to provide coordinated cleft care
- Early intervention to minimize the impact of the cleft
- Outcome measures to review clinical practice and continually improve cleft care (ACPA, 2007:24-25; WHO, 2002). There is a need for assessment and outcome measures to be locally relevant and
- Creating collaborative groups and a global registry and database on craniofacial anomalies (WHO, 2001b: ix).

The guidelines for best practice in cleft care are available from developed countries. However, as discussed, the application of these guidelines in developing countries is challenging. Strategies to improve cleft care globally need to be planned whilst taking into account the

diversity of systems and contexts. Developing countries will have to take responsibility to develop their local capabilities, to improve and sustain cleft care.

The WHO report (2002: 100) recommends the establishment of national registers and databases for individuals with craniofacial anomalies so that data is readily available for clinical and public health action. Professionals and institutions may use this knowledge for international collaborations to address the gaps in cleft care identified through research. A strategy that could be applied to improve cleft care, despite limited number of available speech-language therapists and audiologists, is the leveraging of other resources (training parents, community health workers and other available resources) in communication development. However, to play such a proactive role speech-language therapists and audiologists require access to contextually appropriate assessment instruments.

## **2.5 SUMMARY**

This chapter described the pervasive impact of a cleft on a developing child and on his/her parents and families. An in-depth review of literature was carried out to describe the communication, speech and language characteristics of children with CL/P. The recommendations for standard of care for young children with CL/P from developed countries, namely team approach to assessments and treatment, early interventions and outcome measures of treatment were discussed. Gaps in the service delivery model for young children with CL/P in developing countries, as well as the challenges to upholding optimal standards of care in developing contexts, were also discussed. Additionally, strategies were identified to overcome

specific barriers to cleft care in developing countries. These were to set up database/national register for individuals with CL/P, and provide an interdisciplinary team approach, with an early communication intervention by the speech-language therapists and audiologists for young children with CL/P.