INTRODUCTION

Craniofacial anomalies can be defined as the conditions that literally encompass all congenital deformities of the cranium and face. More specifically, however the term has come to imply congenital deformities of the head and face that interferes with physical and mental well being. (Marsh and Vannier, 1985)

The exact prevalence of craniofacial anomalies cannot be determined as there are no universal epidemiological studies available due to lack of birth registries and register for entries in many countries. This is especially true for the developing and underdeveloped countries. The prevalence studies are mostly based on cleft lip and or palate which are infact the most commonest. The estimated global prevalence of which is 1 out of every 500-700 births. (WHO, 2004)

The spectrum of craniofacial anomalies is very diverse and the most common conditions include (but not restricted to) Cleft lip and or palate, Craniosynostosis (may be associated with Crouzon’s syndrome, Apert’s syndrome), otomandibular anomalies (Treachers Collins syndrome), CHARGE associations, holoprosencephaly, stickler syndrome, fetal alcohol syndrome etc. (WHO, 2002)

The etiology of the craniofacial anomalies is not very well understood and is thought to be multifactorial including both genetic and environmental factors. Various mutations and polymorphisms in many genes are shown to be associated with increased risk of craniofacial anomalies eg. MSX1, TGFA, TGFB, RARA, BCLX3, MTHFR, PAX9, FGFR1, FGFR2, TCOF1 etc. or chromosomal aberration like Down’s syndrome. (Singh et al, 2012;Singh et al, 2011; Wong and Hagg, 2004). Other studies implicit environmental factors such as nutritional factors eg. Vitamins, especially folic acid deficiency in pregnancy is believed to be related with high risk of craniofacial anomalies. Certain agents called as teratogens including various drugs and infective agents are known to induce abnormal embryonic development and may lead to the development of craniofacial anomalies eg. medications like aspirin and valproic acid can induce.
abnormal craniofacial development. Viral infections like rubella can also induce abnormal embryonic development. Similarly maternal smoking and alcohol consumption can lead abnormal craniofacial development. (Wong and Hagg, 2004; Murray, 2002)

The clinical features include a spectrum of deformities of the craniofacial region including cranium and cranial sutures, deformity of skull shape, facial bones including maxilla, mandible and zygomatic arches, nose, eyes, ears, lips and teeth. Along with this there can be systemic involvement including other systems. (Husu et al, 2013; Stavropoulos et al, 2012; Tuna et al, 2011; Kini et al, 2010; Chong et al, 2008; Itthagarun et al, 2007; Lu et al 2007;)

“What is beautiful is good” phrase depicts the importance of facial attractiveness in the social outlook. The importance of physical attractiveness can be estimated by the ever growing cosmetic industry and overemphasis on cosmetic products, exercise and diet products, fashionable clothing and even plastic surgery. We usually perceive attractive people positively. Attractive people are thought to achieve more in terms of various social attributes like employment, personal life and social attention. (Little et al, 2006) Studies even show that in criminal cases the juries have given less severe punishment to the physically attractive defendant. Surprisingly this is true for even children where attractive children are biased by the teachers and peers. (Boyce, 1979; De Sousa et al, 2009; Rosser et al, 2010)

The patients with abnormal facial appearance often have to face discrimination in the society; individuals with abnormal facial appearance are considered to be less attractive and are often considered as less capable, less intelligent and less honest. They face interferences with personal life, employability and social interaction. (Van den Elzen et al, 2012). Many investigations have shown that these disfiguring conditions can lead to various psychosocial problems such as high level of social anxiety, social avoidance and affect the quality of life. (Moss, 2005; Versenal et al, 2010; Van den Elzen et al, 2012).

Craniofacial anomalies comprises of very complex and diverse conditions and they form a considerable proportion of the population. There spectrum is very large; however cleft lip/palate, craniosynostosis forms the major groups. These anomalies have an impact on speech, facial
appearance, hearing and cognition that leads to adverse influences on health and social and emotional integration. (WHO, 2002). Subjects that are affected by craniofacial anomalies (cleft lip/palate) are more dissatisfied with their facial appearance than others and this may be associated with psychosocial functioning leading to an increased risk of social and adjustment problems.

The appearance problems in this patient group are further compounded by an increased prevalence of dental anomalies and malocclusion. (Desilva et al, 2006; Menezes & Vieira, 2008; Da silva et al, 2008; Vettore & Sousa Campose, 2011; Tannure et al, 2012). The treatment approach is multi disciplinary which needs team work and support from various specialty. During almost all phases of treatment, dental services are needed and the orthodontics is almost always needed from early treatment till late adult life. (Lorenzzoni, et al, 2010) Treatment needs are usually high in these patients. Dental aesthetics is an important component of facial attractiveness and various studies have shown that some patients have shown significant increase in self confidence pertaining to body image and appearance after undergoing orthodontic therapy. Good dental aesthetics might have a beneficial role on behavioral and self esteem. (Klages et al, 2006; Kenealy et al, 2007; Rappaport et al, 2010). Therefore the objective assessment of the impact of dental aesthetics on subject well being is very important in clinical practice.

Clinicians are expected to produce evidence of the quality of care they deliver. The quality of the care is not only evaluated by the clinical outcomes but also by the expectations and experiences of the patient during the treatment. (William et al, 2005)

Quality of life has become a very important part in assessment of treatment outcomes and relates to the difference between our expectations and our experience. Identifying the patients expectations of treatment and how it affect their life is important in understanding their oral health needs, their level of satisfaction with treatment provided and quality of treatment delivered. (Sayers & Newton, 2006; Duggal R et al, 2010; Barakati, 2011). However for patients with craniofacial anomalies there is little understanding of the perception regarding orthodontic treatment.
The research in this area is more conflictive and suffers various lacunas due to variety of psychometric scales and constructs used and the lack of validity and reliability in the test population, small sample size, no sub categorization of the anomaly and many other methodological errors. (Singh et al, 2012) Investigators have highlighted the need to move towards a “social science model” from a “medical model”. (Strauss & Broder, 1991; Broder, 1997; Speltz & Richman, 1997; Endriga & Kapp Simon, 1999)

This study was undertaken considering the lack of consensus due to various methodological errors regarding psychological adjustment due to altered facial appearance in patients of craniofacial anomalies and utilized the Derriford appearance scale which is specially designed for evaluating psychological adjustment in people with appearance problems. There are no such studies in the Nepali population as per indexed literature and very few studies globally utilizing the Derriford appearance scale. The psychological impact of dental aesthetics in this group of patients and their expectations from orthodontic treatment is still not studied as per indexed literature.

The objectives of this study were:
1) To assess the psychological adjustment in relation to facial appearance in patients with craniofacial anomalies.
2) To assess the psychological impact of dental appearance in patients with craniofacial anomalies.
3) To assess the expectations of this patient group from orthodontic treatment.

The following hypothesis were put forward:
- H1- There is poor psychosocial adjustment in patients with craniofacial anomalies
- H2- There is a psychosocial impact of dental appearance on patients with craniofacial anomalies
- H3- The expectations of these group is high from orthodontic treatment