

# International Encyclopedia of Rehabilitation

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# **Quality of life among children, adolescents and adults with orofacial clefts**

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## **Introduction**

The term “orofacial cleft” (OFC) covers a wide range of disorders affecting the lips and oral cavity. Generally, one distinguishes between cleft lip (CL; ICD-10: Q36.-), cleft lip and palate (CLP; ICD-10: Q37.-) and cleft palate (CP; ICD-10: Q35.-). Additionally, OFCs may be left- or right-sided (unilateral), or both sided (bilateral).

Orofacial clefts (OFC) are the most frequent birth defects worldwide. One in five- to seven hundred newborns suffers from OFC (Mossey et al. 2009; Eurocat, 2010). Prevalence rates vary as a function of ethnic, geographical, and socio-economic factors, and mothers’ general health status.

## **Aetiology**

Several factors seem to underlie OFCs, though no clear-cut factors have been isolated so far. On the other hand, naïve theories abound. To illustrate, the ancient Britons claimed that a pregnant woman who ate hare's flesh would bear a child with a hare-lip (Thomas, 1983). And these days, Nigerian ethnic groups attribute the occurrence of OFC to an act of God, witchcraft or evil spirits. As a result, only 40% of Nigerian respondents were aware that surgery was an applicable treatment, and only 22% recommended a visit to a clinic (Oginni et al. 2010).

Orofacial clefts result from a failure during embryonic development. Specifically, between the fourth and seventeenth weeks of embryonic development, the closure mechanisms for the facial and oral cavity fail. Note that at the very beginning of embryonic development, the embryo is particularly sensitive to adverse substances, and that women are not always aware of being pregnant at this early stage of pregnancy.

Scientific research focuses on genetics (1), environment (2), and gene-environment-interaction (3).

## **Genetics**

OFC occurs more frequently in monozygotic twins (that is: twins developed from the identical zygote [cell]) than in dizygotic twins. Males are more affected by CL than females. Left-sided clefts are more frequent than right-sided or both-sided clefts. The occurrence of OFC is greater in Latin America and Asia while it occurs less often in Southern Europe, South Africa, and Israel, compared to other countries of the world, whereas the birth prevalence of CL/P in Hamedan city (Iran) was closer to the prevalence of CL/P in the US and Europe than Africa and the Far East (Zandi & Haidari, in press). These observations are consistent with the notion that genetic factors do influence the occurrence of OFC (cf. Mossey et al. 2009).

## **Environment**

Environmental factors include: the mother's unfavourable health behavior such as consumption of tobacco, alcohol, drugs, or medicaments; deficiency of vitamins, zinc and, probably, folate. Comparing demographic characteristics and outcomes for children with OFC in three different states of the USA, Damiano et al. (2009) found that, compared to New York and Iowa, children in Arkansas were born into families with lower income and to parents less likely to be married. Children in Arkansas were also more likely to have special health care needs and require mental health care.

## **Gene-environment interaction**

Additionally, it is conceivable that there are interactions between genetic and environmental factors. To illustrate, the risk of bearing a child with OFC is likely to be increased for women with an observed family history of OFC and concomitant consumption of tobacco and alcohol, as well as with an unplanned pregnancy and deficiencies in vitamins and zinc. A pattern of polycausality has been confirmed in a study performed in Iran (Zandi & Heidari, in press). Note that all these factors do increase the probability of the occurrence, though this does not imply a linear link between OFC, genes and environment.

## **The relation between quality of life, sociopsychological functioning and orofacial clefts**

Generally, facial appearance is regarded as an important prerequisite for healthy psychosocial development (Cole, 1998; cf. Rumsey & Harcourt, 2005). This assumption is supported from three directions. First, evolutionary theories claim that symmetric faces imply physical health, including a well-functioning immune system, the absence of illness or a relative lack of environmental insults during development (cf. Buss, 2008). Second, these theories emphasize that the beauty and attractiveness may depend upon the face's symmetry and a relatively large interocular distance. Third, social psychologists claim that social interactions largely rely upon perception and interpretation of facial expressions (Hubbard, 2001; Nummenmaa et al. 2008; Vigil, 2009) and evaluations of attractiveness (cf. Feingold, 1992).

As a consequence, it might be anticipated that children, adolescents, and adults with untreated orofacial clefts would be at risk for psychosocial difficulties. In this view, untreated OFC generally leads to negative effects on speech, hearing, appearance, and physical and psychological well-being. To illustrate, Akinbami (2007) reported a 25-year-old single female patient with hypernasality and misarticulation of speech secondary to an untreated OFC. The young lady described having problems interacting with other people and with young males in particular.

## **Quality of life among children, adolescents and adults with orofacial clefts**

With regard to the relation between surgically repaired OFC and psychosocial difficulties, results from a large body of research have thus far failed to provide a coherent picture. The reasons for this incoherent picture remain unclear, however, methodological issues may confer to this incoherence: To name a few, issues are related to studies using different measures, non-representative and/or small samples, lack of control groups, lack of longitudinal studies, cultural differences (leading to different degrees of acceptance of visible differences), differences related to the quality of treatment, as well as different registration systems across countries.

## **Quality of life among children and adolescents with orofacial clefts**

In children and adolescents, on the one hand, findings are of a negligible impact of CLP on an individual's psychological well-being (Bressman et al. 1999; Gussy & Kilpatrick, 2006), supporting

the conclusion that in general, children and adolescents might not suffer from psychopathological difficulties (Hunt et al. 2005). Sagheri et al. (2009) investigated 61 children with CLP between four and seven years. The main results were that, compared to children without CLP, children with CLP revealed no statistically significant differences in physical well-being, emotional well-being, self-esteem, family life, or friendships at school.

Similarly, Brand et al. (2009) investigated 32 children and adolescents aged 6 to 16 years with CLP and 34 controls. The main results were that participants with and without CLP did not differ with respect to emotional problems, conduct problems, or hyperactivity. Moreover, with respect to sleep, poor sleep patterns were associated with psychosocial strain, but not with the presence of CLP.

Kramer et al. (2009) reported a very differentiated picture of psychological and family functioning in children with OFC. They investigated 170 children from 8 to 12 years with and without OFC and found that family functioning was superior in families with children with cleft lip only (CL), compared to families with children with cleft palate (CP) only or cleft lip and palate (CLP). Most importantly, general quality of life was superior in children with OFC compared to controls without OFC. Problems in the social field such as how to behave with family members or friends were specifically reported by children with CLP. Kramer et al. (2009) indicated therefore the need for research to distinguish between the different types of OFC.

In the same vein, Feragen et al. (2009) reported that in children aged 10 visibility of the cleft, gender, and additional diagnoses had no negative impact on psychosocial resilience, that is, the positive capacity to cope successfully with psychosocial issues (cf. Baker et al. 2009; Feragen et al. in press). Likewise, Snyder and Pope (2010) also found that some children and adolescents with craniofacial anomalies reported lower rates of problems compared to norms. Snyder and Pope (2010) interpreted this finding as evidence that children and adolescents might build up a positive capacity to successfully cope with psychosocial issues. Additionally, in a Chinese sample, scores for satisfaction with life and social anxiety of people with OFC did not differ from controls (Cheung et al. 2007).

Last, to illustrate the complexity of social perception and social interactions, Edwards et al (in press) could demonstrate that first impressions others have of youth with clefts were significantly affected by how these youth present themselves in social situations. As a result, the presence of a repaired cleft might be more or less perceived and interpreted as irritating as a function of positive or unfavorable social skills of a person with OFC.

On the other hand, there is also a large body of research which shows that children and adolescents with OFC suffer from specific socio-psychological issues. Brand et al (2009) showed that children and adolescents with CLP were six times more likely to report difficulties in interactional competencies compared to controls without CLP. Stigma experiences, social problems (e.g., Snyder et al. 2005), symptoms of anxiety and depression, dissatisfaction with facial appearance (e.g., Patrick et al. 2007), withdrawal and internalizing problems (Pope & Snyder, 2005), and shyness (Boes et al. 2007) have also been reported.

In a cross-sectional study, Hunt et al. (2006) assessed the psychological functioning of patients with CLP, comparing 160 children and young adults with an age- and gender-matched control group. The main results were that, compared to controls without CLP, children and young adults with CLP reported greater behavioral problems, increased symptoms of depression, and a lower satisfaction with facial appearance and speech. Moreover, these participants indicated that they were more likely to be teased in social settings. No significant differences between participants with and without CLP were found for anxiety and self-esteem. However, independently of the presence of CLP, having been teased predicted significantly poorer psychological functioning. Hunt et al.

(2006) concluded that, as part of routine cleft care, children and young adults with CLP require psychological assessment, specifically focusing on their experiences being teased.

Feragen et al. (2009) reported that cleft types were related to the frequency of additional physical and psychological difficulties, pointing to a need to differentiate between types of OFC. They were also able to show that psychosocial resilience was related to adequate emotional functioning, high satisfaction with appearance, and low reported frequency of being teased. Last, compared to children and adolescents with other than OFC, those with OFC registered higher scores for social problems and deficits in social, and academic competencies. Moreover, adjustment seemed to be related to appearance and speech deficits, which in turn appeared to be associated with poor peer relationships. Specifically, poor adjustment was observed in those children reporting increased teasing by peers (Snyder & Pope, 2010; see also Wehby & Cassell, 2010).

Taken together, for children and adolescents, the occurrence of a repaired OFC does selectively negatively impact on quality of life. Specifically, compared to healthy controls and normative data, specific social interactions seem to be affected. With respect to peer relationships, children and adolescents indicate that teasing is particularly unpleasant. However, there is also evidence that the presence of a repaired OFC does not necessarily lead to unfavourable psychosocial outcomes; on the contrary, OFC can also be associated with increased resilience.

### **Quality of life among and adults with orofacial clefts**

As for children and adolescents, results related to the quality of life of adults with orofacial clefts are inconsistent.

Cheung et al. (2007) found that among Chinese adults with OFC, compared to controls, social anxiety was not higher, but self-esteem was lower. In the same vein, Berk et al. (2001) reported that, compared to siblings without OFC, adults Chinese with OFC had poorer scores for self-esteem and social support and, additionally, higher scores for social anxiety. Berk et al. (2001) concluded that, compared to siblings without OFC, adults with OFC may be more disadvantaged with respect to social affiliation and adaptation.

In contrast, Chetpakdeechit et al. (2009) found that young adults with OFC and receiving recognition from significant others reported higher self-esteem and greater ability to cope with their social lives. Ramstad et al. (1995b) found that, compared to healthy controls, adults with OFC reported anxiety and depression about twice as often, and these psychological problems were strongly associated with concerns about appearance, dentition, speech, and desire for further treatment. To summarize, these findings point to potential difficulties in psychological wellbeing among some adults with clefts.

Marcusson et al. (2001) investigated a Swedish sample of 68 adults with OFC and 66 adults without OFC. Compared to the gender- and aged-matched control group, adults with OFC had significantly lower scores for quality of life, family life, private economy, global life, disturbance to life, well-being, and social contacts. However, more practical and tangible aspects of the daily lives of the adults with OFC were not affected. Marcusson et al. (2001) concluded that adults with OFC reported a fairly good life adjustment in spite of the presence of the facial malformation.

Danino et al. (2005) investigated 82 French adults with repaired CLP and found that, compared to adults without CLP, people with CLP did marry later, displayed a delay in scholarship, had a lower income, and reported a significant delay in their independence process from their parents. These findings do match those based on observations of a Norwegian sample of 233 adults aged 20 to 35 with repaired OFC: Compared to the general population, adults with OFC were less likely to marry and, if they did so, to marry later in life, giving birth to fewer children (Ramstad et al. 1995a).

However, data from a Norwegian population-based cohort study did suggest that any kind of birth defect, including OFC, was not unfavourably associated with adult intellectual performance, disability and mortality. Thus, it seemed that the presence of a repaired OFC had no negative impact (Eide et al. 2006). Patel and Ross (2003) investigated 20 South African adults with repaired OFC and concluded that participants were generally satisfied with their current communicative abilities and with their educational attainments. Employment did not appear to be affected. As children, they had been teased, though they also always had good relationships with their teachers. Moreover, as adults, dating and marital relationships did not seem to be adversely affected. Last, Mani et al. (in press) investigated 86 Swedish adults with OFC and showed that compared to older participants, younger participants were more negatively affected in several areas related to quality of life such as social, physical and emotional role function. However, when compared to normative data, participants with OFC did not significantly differ from norm data.

Taken together, as for children and adolescents, some aspects of quality of life of adults with OFC may be affected. However, findings vary with assessment tools, normative data and study focus. Most importantly, the occurrence of repaired OFC does not necessarily lead to unfavourable psychosocial outcomes.

### **Scientific journals (selection)**

- British Journal of Oral Maxillofacial Surgery
- European Journal of Orthodontics
- International Journal of Oral and Maxillofacial Surgery
- Journal of Cranio-Maxillofacial Surgery
- The Cleft Palate Craniofacial Journal
- The Journal of Craniofacial Surgery

### **Internet sites related to OFC (selection)**

- <http://www.cleft-children.org/> (multilingual)
- <http://www.cleftnet.de/> (German)
- <http://cleftworld.com/>
- <http://www.clapa.com>

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