Living with Crouzon syndrome: transition from childhood to adulthood

Ulrika Hallberg, Dimitrios Stavropoulos, Bengt Mohlin* and Catharina Hagberg

Nordic School of Public Health, Gothenburg, Sweden; Department of Orthodontics and Jaw Orthopedics, Institute of Odontology, The Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden

(Received 3 March 2010; accepted 8 December 2010)

Objective. The aim of this qualitative investigation is to study the subjective experiences of transition from childhood to adulthood in individuals with Crouzon syndrome. Material and methods. Telephone interviews were carried out with eight informants and data were analysed according to grounded theory. Results. A core category emerged labelled facing barriers when developing self-image, which illuminates the different barriers the child had to face when trying to develop a self-image during the transition from childhood to adulthood. Facing barriers should not be confused with the actual barriered development. These barriers are further illuminated in five descriptive categories. Conclusions. These children face a variety of stressful barriers when developing their self-image during the transition from childhood to adulthood. The produced psychological outcome seems to be related to a complex interaction among multiple variables, including variables related to the individual with Crouzon syndrome, treatment variables, parent and family variables and social variables.

Keywords: adolescence; adulthood; childhood; Crouzon syndrome; grounded theory

Introduction

Crouzon syndrome is a congenital developmental deformity syndrome, mainly of the cranial skeleton, with a birth prevalence of approximately 16.5 cases per million live births (Cohen and Kreiborg 1992). In half of the cases, the syndrome is caused by a new mutation. Otherwise, the syndrome has an autosomal dominant inheritance. The gene is localized on chromosome 10. It is included in the clinical entity of craniosynostoses, a heterogeneous group of syndromes characterized by premature fusion of cranial sutures, giving rise to multiple anomalies of the growing and developing craniofacial region (Kreiborg 1986). Typically, the resultant cranial disfigurement is characterized by brachycephaly, frontal bossing and low hair line. Exophthalmos, hypertelorism, parrot-beaked nose, short and retracted upper lip, maxillary hypoplasia and dental malocclusion (open bite, posterior cross bite, Angle class III malocclusion, severe dental crowding) are the cardinal clinical features of the dentofacial disfigurements. Main clinical symptoms include neurological symptoms (headaches in 25%, seizures in 10%, mental retardation in 15%), ophthalmologic symptoms (divergent strabismus or exotropia in 75%), exposure

*Corresponding author. Email: mohlin@odontologi.gu.se
conjunctivitis or keratitis in 50%, blindness in 7%), otolaryngologic symptoms (hearing loss in about 50%) and orthopaedic symptoms (stiffness of joints, especially the elbows) (Kreiborg 1981).

Patients with Crouzon syndrome require a multidisciplinary treatment approach, involving, among others, several surgical interventions over time, usually from the first months of life to improve function and aesthetics (Posnick and Ruiz 2000). Nevertheless, a functionally and cosmetically good result is often hard to attain and psychological distress comes as an unavoidable consequence, due to the various kinds of stigmata persisting in the craniofacial region.

Physical appearance concerns are at a peak during adolescence, therefore, teenagers with facial differences may have challenging experiences at this particular time (Charkins 1996). This is also a time when young people place considerable emphasis on social relationships (Bradford, Rutherford, and John 2002). Edwards et al. (2002), in their study on adolescent quality of life, found that ‘friends’ and ‘belief in self’ were of the most frequently cited themes in adolescent interviews.

Facial appearance is immediately observable and a visible facial difference is quickly perceived in social discourse (Macgregor 1979). Research on children with craniofacial anomalies indicates that dissatisfaction with facial appearance is related to greater loneliness, fewer same-sex close friends, social withdrawal and dislike by peers (Pope and Ward 1997). Furthermore, adolescence is a unique developmental period, in which social and media norms frame self and peer expectations and play a significant role in creating and exacerbating pressures on those distressed by their appearance. The pressure caused by the barrage of images of ‘beautiful people’ in programs and articles which encourage as to critically evaluate our appearance and to correct the ‘faults’ may be more keenly felt by those who have little hope of conforming to the unrealistic ‘norms’ manufactured by the media (Rumsey and Harcourt 2004). Facially disfigured children frequently experience traumatically offensive remarks, unpleasant stares, stunned reactions and outright avoidance (Macgregor 1990). Similar reactions in young adults treated due to cleft lip and palate (CLP) were found in a recent qualitative study (Chetpakdeechit et al. 2009). The core category in that study was ‘Hoping to be like other people’. Most of the subjects had received extensive treatment but the individuals still retained a feeling of being different. Treatment in order to improve appearance could even contribute to strengthen these feelings. Professional evaluations have often been focused on the morphological results of treatment. The optimal balance between the amount of treatment and the creation of a reasonable acceptable appearance is difficult to establish. Some of those with CLP stated that they still as young adults were subjected to teasing due to deviating appearance. Responses from others can produce a strong effect on self-concept, because the self-esteem system assesses the extent to which one is accepted or rejected by others (Leary and Downs 1995). Consequently, children with craniofacial anomalies may be at a greater risk for negative self-evaluations of appearance and reduced overall sense of self-worth (Broder, Smith, and Strauss 1994; Maris et al. 1999). Pruzinsky (1992) summarizes four potential problem areas in children with a major craniofacial deformity: low cognitive development, negative emotional attachment between the child and parents, impaired development of peer-relations and the experience of shame.

Studies of the life situation of people with a facial malformation are often inhomogeneous, in that the study groups include different syndromes of congenital or even acquired malformations. This is because of the low prevalence of syndromes.
However, there are obvious limitations in internal and external validity issues. Therefore, the aim of this qualitative investigation is to study the subjective experiences during transition from childhood to adulthood in individuals with Crouzon syndrome.

Material and methods

Study group and procedure

An informational letter about the study was sent out to 19 young adults with Crouzon syndrome (identified from patient records from the jaw orthopaedic unit of the Gothenburg university clinic of orthodontics), living all over Sweden, asking them if they were willing to participate in the study. If they replied positively they were asked to contact one of the authors (UH, sociologist and doctor of public health science) to schedule a time for a telephone interview. The interviewer was neither a member of the clinical care staff nor known to the participants in advance. A telephone interview lasting up to 90 minutes was conducted with each one of the participants in the study. The sample consisted of six males and two females, with Caucasian ethnic background, aged from 18 to 37 years (mean age 25.4 years). They had variable severity of the syndrome and stated that they had undergone many craniofacial surgical operations, starting from their childhood years. Background data of the study sample are presented in Table 1. The interviews were held in a conversational style. An interview guide was used and covered themes such as childhood, school situation and relations with friends and family. The participants had the opportunity to raise subjectively important questions regarding the area under study and the interviewer asked relevant follow-up and probing questions. Data were generated within the process of active involvement of both the researcher and the informant in responding, clarifying and elaborating communication.

Data collection and analysis were performed using a qualitative research method. In areas with limited knowledge such methodology has the ability to grasp the participant’s point of view, which among other things, may create a better base for further quantitative studies (Hallberg 2006). Qualitative research can be defined as a method involving the systematic collection, organization and interpretation of textual material derived from talk or observation (Malterud 2001). Such methodology can be used in the exploration of meanings of social phenomena as experienced by individuals themselves in their natural context. The qualitative method applied in the present study was the grounded theory, as was originally described by the two

<table>
<thead>
<tr>
<th>Participant</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Employment status</th>
<th>Residence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25</td>
<td>Male</td>
<td>Unemployed for a year</td>
<td>Lives with parents</td>
</tr>
<tr>
<td>2</td>
<td>26</td>
<td>Female</td>
<td>Master degree student</td>
<td>Lives in own apartment</td>
</tr>
<tr>
<td>3</td>
<td>21</td>
<td>Female</td>
<td>Teacher student</td>
<td>Lives with parents</td>
</tr>
<tr>
<td>4</td>
<td>25</td>
<td>Male</td>
<td>Sick leave for 2 years</td>
<td>Lives with parents</td>
</tr>
<tr>
<td>5</td>
<td>24</td>
<td>Male</td>
<td>University student</td>
<td>Lives in own apartment</td>
</tr>
<tr>
<td>6</td>
<td>18</td>
<td>Male</td>
<td>High school student</td>
<td>Lives with parents</td>
</tr>
<tr>
<td>7</td>
<td>37</td>
<td>Male</td>
<td>Runs his own company</td>
<td>Lives in own apartment</td>
</tr>
<tr>
<td>8</td>
<td>25</td>
<td>Male</td>
<td>No answer</td>
<td>No answer</td>
</tr>
</tbody>
</table>
sociologists Glaser and Strauss (1967) in the 1960s and advanced by Strauss and Corbin (1998), and later by Charmaz (2000). Grounded theory, which places an emphasis on social dynamics, has its theoretical roots in the sociological theory of symbolic interactionism, bearing the assumption that people construct meanings about their lives on the basis of interactions they have with other people and the world at large (Blumer 1969). Grounded theory is an inductive research method, which aims at generating concepts, models or theories developed from empirical data (i.e., grounded in data), in order to explain and/or predict the phenomenon under study.

The basic principles of grounded theory include constant comparisons, theoretical sampling, saturation and theoretical sensitivity. Constant comparisons include that one piece of data is being constantly compared with other pieces of data. Concepts must earn their way into the data (Glaser 1992) and therefore, different parts of the data are continuously compared in terms of differences and similarities. Also, a specific category is compared with other categories, as well as data from different subjects and new data are being compared with an emerging category. The emerging categories guide further questioning, that is, theoretical sampling. The theoretical sampling procedure aims at refining theoretical ideas and saturating emerging categories. Saturation, although somewhat ‘elastic’, is reached when new interviews bring no additional information, that is, new data fit into the categories already devised (Charmaz 2000). Theoretical sensitivity refers to the researcher’s reflexive way of developing research questions and doing analysis (Hall and Callery 2001).

In the present study, open audio tape-recorded interviews were conducted with the informants. Data collection continued until nothing new emerged in the analysis. Nevertheless, since the possible participants of the study were few (n = 8), the reached saturation was judged to be tentative.

The audio tape-recorded interviews were transcribed into text (verbatim transcribed interviews) and analysed in open, axial (theoretical) and selective coding processes (Glaser and Strauss 1967; Strauss and Corbin 1998). Open coding means that through a line-by-line reading of the transcribed into text interviews, the substance of the data was caught and segmented into substantive codes, which were labelled concretely. The process of open coding ended up with clustering substantive codes with similar content into summarizing categories. These categories were given more abstract labels than the substantive codes belonging to it. In the axial (theoretical) coding, each category was further developed. Relationships between categories were sought and data were put together into a new wholeness. In the selective coding process, the categories were saturated by additional information, assessed by new interviews or added by re-coding earlier assessed data. A core category was identified, describing the main concern for adolescents with Crouzon syndrome. This core category was central in the data and could be related to all other categories. During the entire process of analysis, ideas, preliminary assumptions and theoretical reflections were written down in notes or ‘memos’.

The study was approved by the Regional Research Ethics Committee of Gothenburg (registration number Ö 342–99).
Results

In the data analysis, a social process emerged describing the transition from childhood to adulthood in children with Crouzon syndrome and was reflected in the core category, which was labelled: facing barriers when developing self-image. This illuminates the different barriers the child had to face when trying to develop the self-image during the transition from childhood to adulthood. These barriers are further analysed in the following descriptive categories, labelled: being aware of differences in the self, being aware of deviation in facial appearance, being regarded as different by others, having to limit the self according to others, and going through identity crises (Figure 1). It is not infrequent that even individuals with clear medical records often have to go through and cope with various social barriers, when developing the self-image during the transition from childhood to adulthood. However, in people with Crouzon syndrome, these barriers are more extreme and the individual is more vulnerable and exposed due to the syndrome. The categories are probably mainly to be seen as context specific. Individuals with severe health problems like mental retardation and severe hearing problems were not able to participate. Medical interventions in order to reduce complications related to craniosynostoses are started at an early age. It is not likely that the subjects remember these interventions. The very obvious and characteristic craniofacial deformities are likely to be the main reason behind the barriers experienced by the informants.

Short, exemplary quotations from the interviews, translated from Swedish, are presented below the results obtained, to further illuminate the descriptive categories.

Figure 1. The relationship between the core category of concern for Crouzon syndrome individuals and the descriptive categories identified from the telephone interviews.
Facing barriers when developing self-image
During very early childhood, it seems that children with Crouzon syndrome are not aware of their condition and feel like everybody else. When they start to socialize with other children on a daily basis, they become aware of differences in themselves compared to others. Children with Crouzon syndrome have to visit medical care units frequently and they are, accordingly, frequently absent from daycare centres, school and friends. This can be described as a barrier for developing the self for the young child, followed by the awareness of facial difference, which constitutes a new barrier. The child experiences others regarding him/her as different and he/she is often exposed to bullying. Furthermore, the child is exposed to identity crises, owing to facial changes due to craniofacial plastic and reconstructive surgery. Additionally, he/she experiences feelings of being limited in his/her self-image development by overprotective parents. The child with Crouzon syndrome has to face and cope with all these barriers in order to develop the self-image during the transition from childhood to adulthood.

Being aware of differences in the self
Even from an early age, children with Crouzon syndrome are obliged to attend several hospital visits for medical examinations and treatment. This is something most of their peers do not have to experience. When the child with Crouzon syndrome becomes aware of this, he/she feels different from others, not necessarily in a negative way. The child, not fully aware of the reason for the frequent hospital visits and, being separated from its friends, just accepts the fact and complies with the parents’ wish. The child is not, at this point, aware of deviations in his/her facial appearance. According to the participants of the present study, they did not think about their appearance at all at that early age. Several of the early interventions were accomplished due to other reasons than concern for deviating appearance.

‘... of course, I was well aware that I was different to some degree, since I spent a lot of time at the doctor’s, you know ...’

Being aware of deviations in facial appearance
During their first year in school, or directly before they started school, the informants became aware of deviations in their facial appearance. This was because their friends or schoolmates asked them about their facial appearance and why it differed from others. This made them start thinking about their face and they compared themselves to their peers. During this comparison they realized their facial disfigurement. This awareness was not necessarily negative for them. It could just be considered a fact that they looked different from others and became curious why it was so. The participants of the study described that they asked their parents about their facial appearance and how their parents tried to explain to them what was unique with them and that they were born in that way.

‘There was a girl at preschool that asked me why I looked different. That’s when I became aware of my appearance. She just asked, it wasn’t anything negative, it was just out of curiosity.’
Being regarded as different by others

The participants of the study also described that their schoolmates regarded them frequently as different from others. This was sometimes experienced to have a negative influence. They spoke about harassments from others, both from their peers and from strangers they met. They were called names and they were rarely allowed to play with other groups of children. This made them feel lonely and vulnerable. In some instances, the children with Crouzon syndrome had just one or two very close friends they could trust and rely on. These peers were the ones who stood up for them, when being bullied. If the informants’ parents supported them and visited the school premises early in their childhood, the harassments could be mediated. This was attributed to explanations given to the other school children about the Crouzon syndrome situation. Nevertheless, the insulting comments could go on into the late teens. Such harassments could be highly harmful, particularly when it occurred in public places. In such places, the children with Crouzon syndrome could be stared at, pointed at, or even asked about their face by indiscreet strangers. This was described as a situation that was scary and difficult to overcome, because the informants were unable to give in advance information about their syndrome to any unknown person they met.

‘When you were little it didn’t matter, it wasn’t the most important thing then, except... but when you started seventh grade, and didn’t really know what it was all about, of course you were teased and excluded and things like that... you weren’t included, that’s what happened then...’

Having to limit the self according to others

The study’s participants described how their parents overprotected them. It could be that they were not allowed to play rough games as other children did, or that their parents watched them continuously and limited their responsibilities, such as going alone to the store, or going too far away from the home. This overprotection was experienced as a situation that hindered them from fully developing themselves. They felt limited and passive. Instead, they wanted their parents to treat them like everyone else, to allow them to play and to climb trees and to have responsibilities like other children at their age had. The more overprotected they felt, the less initiative they took in exploring and taking control over their lives and the more anxious they felt when meeting new children or adults.

‘So, even if I tried to manage on my own, I always felt watched and I still felt very protected by the people I was with... um, it was often like, you know, that they never allowed me to do anything that could subject me to danger or something... um, they never let me try anything.’

Going through identity crises

In order to minimize their facial disfigurement and treat the Crouzon syndrome related health problems, the informants had to go through various surgical procedures, both minor and major, with the major ones taking place mostly during their teens. These operations were physically painful and difficult to cope with, especially during childhood, when the child had limited knowledge of what to expect. These surgical reconstructions also led to a more or less changed face. Even if this
resulted in an appearance that deviated less from the ‘norm’, it was a departure from before, which could lead to identity crises, especially during adolescence. Being used to their former facial appearance and having structured an identity from it, they now had to form a new one based on their ‘new’ facial appearance. These identity crises were experienced as hard to cope with and led to feelings of insecurity and anxiety, when the teenagers with Crouzon syndrome returned to their school presenting their new face.

‘Then, as a teenager, it became maybe even more overwhelming, since you changed the way you looked every other year during that period . . . it was a little, well . . . hormones play a part and you become a little, you know . . . emotionally, it can be very difficult and things like that.’

Discussion

The main finding of the present study, as denoted on the core category, is that children with Crouzon syndrome face barriers, which they have to cope with, when trying to develop their self-image during their transition from childhood to adulthood. This has also been emphasized by Pruzinsky (1992), who notes that children with severe forms of craniofacial deformities experience many psychological stressors to which they must learn to adapt and may potentially affect the children’s psychological development. Similar observations have also been made on children having a chronic physical illness. Wallander et al. (1988) reported that these children have to face stressors, which include such factors as repeated clinical evaluations throughout childhood, multiple and lengthy hospitalizations, and visible differences in physical appearance. They concluded that children with chronic physical disorders were perceived as evidencing on the average, more behavioural and social competence problems than the expected based on norms for children in general.

Growing and developing with a facial disfigurement is regarded as an adverse situation, partly because during the transition from childhood to adulthood the self is still under constant development. Self-concept formation is based on the feedback an individual receives from others (Jourard 1963). Goffman (1963) describes how an individual having a visible facial difference may suffer from stigma either enacted or perceived. Enacted stigma occurs when the individual directly experiences the adverse effects of stigma, implying that others have treated the individual negatively, as in teasing. Perceived stigma is predisposed by the individual’s lowered self-esteem and often is the consequence of the internalization of enacted stigma experiences. Perceived stigma can be seen, for instance, in a child who fails to engage others in play with him/her. Stigma may result in shame and self-image shattering, leading to potential detrimental social effects. The stigmatized have to learn to live in a society where they are not considered complete, are looked down on and often segregated.

Feeling different when comparing themselves to others, was expressed in the descriptive categories ‘being aware of differences in the self’, ‘being aware of deviations in the facial appearance’, and ‘being regarded as different by others’. Therefore, it is worthwhile considering the nature of difference and the time when it becomes important as a life determinant in this group of people. To feel different is in line with observations in a similar study carried out in young adults with cleft lip and palate where the core category was labelled ‘hoping to be like other people’ (Chetpakdee-chit et al. 2009). In the present investigation, being frequently absent from day-care units, in order to attend medical visits, was the only distinguishing difference that
was felt during early childhood from the informants with Crouzon syndrome, but not in a negative way. They did not feel segregated. This is in line with research reports that although children as young as three–four years old are able to distinguish a marked visible difference (Langlois and Styczynski 1979), they do not express stigmatizing behaviours, as it is implied by Kruckeberg et al. (1993), who found that preschool-age children (three–six years old) with various craniofacial anomalies seemed to be more similar than different, compared to a matched control group of healthy children. However, children who were profoundly deaf, or with a low cognitive/mental development index, were excluded from the research groups of the latter study.

It has been discussed that it is during the first years of school that children make judgements about physical attractiveness in peers, which look like adult perspectives (Bull and Rumsey 1988). It is when children begin to differentiate based upon appearance. It has been documented that it is around seven–eight years when children's own preferences (what they like) affect their social cognition (Kalish and Shiverick 2004); it is this age when they start to form stereotypes and show stigmatizing behaviours (Hearst 2007). In fact, there is evidence that this process may start even earlier (Miller and Aloise 1989). Eder (1995) reports that in comparison to healthy children, five-year olds with craniofacial deformities felt more alienated by others, more aggressive, more scared, mad and upset.

Insulting comments and exclusion from children play-groups were frequent behaviours that were expressed on the participants in our study, during their school-years. They were also confronted with repetitive bullying. This is defined as conscious, repeated acts of physical, verbal, or relational aggression that causes injury or discomfort to the target (Olweus 1994), with a more powerful attacking a less powerful one (Nansel et al. 2001). It is most likely to occur in the school playground, manifesting at a peak at seven–eight years (Whitney and Smith 1993), or at 11–13 years, according to others (Nansel et al. 2001). The transition from infant to primary and primary to secondary school, is particularly difficult for children with a visible difference, as they move to successively larger and less protective environments; there, questioning, teasing, bullying and social ostracism may be harder to control (Hearst 2007). The whole situation seems to continue into the late teenage years, a period of development often dominated by self-doubt and insecurity for almost every young person. Thus, having a visible facial deviation during this period of life, when adolescents strive to acquire a sense of personal identity (including acceptance of appearance), an individual value system and social conscience, may present particular challenges; this is because the physical and psychological changes associated with adolescence increase the focus on physical appearance for the majority of adolescents (Rumsey and Harcourt 2004). A qualitative study of young adults with cleft lip and palate gave similar results. Some of these individuals had experienced bullying also as adults (Chetpakdeechit et al. 2009).

An additional impediment that children with Crouzon syndrome have to face during their childhood is parental overprotection, as described in the category of the present study ‘having to limit the self according to others’. ‘Belief in self’ and ‘friends’ has previously been found to be the most cited themes in studies of adolescents (Edwards et al. 2002). The central role of these items seems to be confirmed in the present study. While protecting one’s young from the dangers of the outside world is normative, overprotection suggests behaviours beyond what most parents would do in similar circumstances (Thomasgard and Metz 1993). It has been described as a
common consequence of the emotional turbulence surrounding the birth and care of a child with a craniofacial anomaly (Speltz et al. 1994). Although well-intentioned, the parent may hamper the child’s drive for autonomy and self-actualization, by becoming over-controlling and intrusive in interactions with the child (Mahoney 1988). This may result in affecting negatively the child’s self-perception and undermining of its social skills (Speltz et al. 1994).

The development of a coherent and organized sense of identity is a key task in adolescence (Erikson 1950). Central to the identity formation process is the idea of crisis and the exploration of possible selves and roles, enabling young individuals to create adult selves and to assume adult responsibilities. Such episodes of crises were experienced as intense and hard to cope with by the informants with Crouzon syndrome of the present study. This was attributed partly to the sudden and often dramatic changes produced by the surgical facial reconstructions, which placed immediate demands on the patients’ adaptive skills. This is consistent with observations that when a young person is trying to integrate his/her facial appearance into a stable identity and overall body image scheme, this is even harder to achieve at a time when facial appearance is likely to undergo radical change with surgery; the old face, however imperfect and disliked, is still the more familiar one and its loss needs to be recognized and even mourned (Hearst 2007). Research on orthognathic surgery patients indicates that it may take up to two years to establish as permanent the effect of facial change, because this time may seem necessary for the personality to accept and incorporate the new features (Kiyak et al. 1984). This remains to be validated for people with Crouzon syndrome.

The transition from childhood to adulthood in individuals with Crouzon syndrome concerns a social process. To date, there is a shortage of reports dealing with the above described situations. According to Glaser and Strauss (1967), grounded theory is especially suitable when studying social processes, as well as when investigating how people handle or behave in a special situation (Glaser 1998). It is a particularly useful research method when there is no existing theory in the field under investigation. In line with the above, the qualitative inductive research method of grounded theory was chosen for the present study.

An assumption in qualitative research that should not be forgotten is that data are generated in the interaction between researcher and informant (Strauss and Corbin 1998; Charmaz 2000). Furthermore, it is important to be aware that a generated theoretical model is the result of an informant’s interpretation of his/her reality, which is then interpreted by the researcher, that is, interpretations of interpretations (Strauss and Corbin 1998).

Telephone interviews were preferred in this investigation due to practical issues, such as work commitment of some of the participants and long distances between the participants and the interviewer. Data in qualitative research can be collected in many different ways. Observations, letters, in depth interviews by telephone are some examples. Although some aspects of non-verbal communication may be missed, telephone interviews could, on the other hand, help in creating a neutral environment for the conversation.

Two more limitations of the study should be taken into account. Firstly, there is the potential for an ascertainment bias in the participants who were recruited into the study. As noted above, individuals with Crouzon syndrome seem to be more shy and reticent than their unaffected peers, therefore, those who volunteered to participate in the present investigation and to tell us about their lives and experiences may be
unrepresentative of their less outgoing peers with Crouzon syndrome. There are also a rather high proportion of hearing problems in these subjects. Some of these individuals are mentally retarded. Crouzon syndrome is an unusual syndrome and as focus in this study was on young adults, the number of available participants for this study was limited, resulting in a somewhat limited saturation. Therefore, the results presented are referred to as tentative results and the study should be seen as a pilot study. Nevertheless, according to Charmaz (2000), the unit of analysis in a grounded theory study concerns events and stories, rather than the participants per se. Hence, the number of informants is less interesting than the content and the quality of the data.

Results from a single qualitative study should not be generalized. Additionally, the findings are only applicable to people living under the same cultural and natural context as the informants. In the present investigation, the interpretation of the results pertains to children with Crouzon syndrome grown up in the south west of Sweden without mental retardation or severe hearing problems. The presentation of the current results should be looked upon as a substantive model for understanding the concepts studied. As mentioned before a major benefit with a qualitative study is to generate ideas with association to social processes and phenomena. This will in turn ease further research to, among other things, formulate relevant questions to be used in the future research.

In conclusion, children with Crouzon syndrome face a variety of stressful barriers when developing their self-image during the transition from childhood to adulthood. The produced psychological outcome seems to be related to a complex interaction among multiple variables, including variables related to the individual with Crouzon syndrome (e.g., cognition, social competence), treatment variables (e.g., therapeutic needs, outcome of habilitation), parent and family variables (e.g., parenting style, social support) and social variables (e.g., definition of ‘normal’ and ‘abnormal’, acceptance of difference). Therefore, strategies addressing the habilitation of this group of people should focus not only on medical and psychological treatment modalities, but also on public health interventions for reducing discrimination and negative adolescent experiences related to facial differences.

Acknowledgements
We would like to thank all the participants in the present study for sharing their thoughts and feelings with us. We also thank the Västra Götaland County Council and the Gothenburg Dental Society for financial contribution to this study. We are also grateful to Ellen Asp for professionally transcribing all the interviews and to Sandra Ståhlberg for translating the verbatim transcribed interviews from Swedish into English quotations.

References


