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Living with Crouzon syndrome: how do young adults with Crouzon syndrome handle their life situation?

DIMITRIOS STAVROPOULOS[1], ULRIKA HALLBERG[2], BENGT MOHLIN[1] & CATHARINA HAGBERG[1,3]

[1]Department of Orthodontics, Institute of Odontology, the Sahlgrenska Academy at the University of Gothenburg, Gothenburg, Sweden, [2]Nordic School of Public Health, Gothenburg, Sweden, and [3]Division of Orthodontics and Paediatric Dentistry, Department of Dental Medicine, Karolinska Institutet, Huddinge, Sweden

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Background. Recent research has been focused on those attributes that appear to buffer a person against the stresses and strains of living with a visible difference.

Aim. To provide some insight on how young adults with Crouzon syndrome handle their life.

Design. Telephone interviews were carried out with eight Crouzon syndrome individuals (six males, two females, mean age 25.4 years) and data were analysed according to the qualitative method of grounded theory.

Results. The informants' main concern was *to make the best of their situation*, showing that even in adverse conditions, as in Crouzon syndrome, several individuals do find ways to live with their difference and to succeed in various aspects of life, using strategies they construct. Such strategies, as identified from the present investigation, were labelled: *committed to an engaging activity*, *avoiding exposed situations*, *actively launching oneself*, *struggling with normalizing facial appearance*, and *lowering the expectations of finding a love partner.*

Conclusions. The adaptation of successful coping strategies seemed to be crucial in the quest of attainment of higher self-esteem. The more the participants in the study used the coping strategies they had developed over time, the better they handled their life situation, which led to enhanced well-being.

Introduction

Although Crouzon syndrome features have been described by several authors, even before 1900, the French neurologist Octave Crouzon is generally credited with the identification of the syndrome. In 1912, he described the hereditary syndrome of *craniofacial dysostosis* in a mother and her son, depicting the characteristic triad of calvarial deformities, facial anomalies, and exopthalmos[1]. Kreiborg[2] noted that the syndrome was not reported under the name of *craniofacial dysostosis* or *Crouzon syndrome* in the English literature until 1939; however, several reports of the condition had been presented under the diagnosis of *oxycephaly*.

Typically, the resultant cranial disfigurement is characterized by brachycephally, frontal bossing, and low hair line. Exophthalmos, hypertelorism, parrot-beaked nose, short and retruded upper lip, maxillary hypoplasia, and dental malocclusion (open bite, Angle's class III occlusion, severe dental crowding) are the main clinical findings of the disfigured face.

A specific trend can be distinguished in the relevant Crouzon syndrome literature. The early studies were primarily oriented towards the neurological and neurosurgical problems; later, the ophthalmologic problems were emphasized, and then problems related to craniofacial plastic surgical reconstruction were focused on. Lastly, the psychosocial issues were addressed, under the recognition of the importance of social competence as a significant component of psychological health[3] and the acknowledgement that individuals with a facial disfigurement can be stigmatized and have difficulties with social situations[4].

Correspondence to:
Dimitrios Stavropoulos, Department of Orthodontics, Athens Naval Hospital, 70, Dinokratous Str, GR – 115 21, Athens, Greece.
E-mail: dimitrios.stavropoulos@odontologi.gu.se

The face is critical and unique in human development and social interactions[5]. A visible difference comprises a 'social disability', as in addition to impacting on the thoughts, feelings, and behaviours of those affected, it is also likely to be noticed by other people; such unwelcome attention and the consequent lack of anonymity in a crowd has been described as visual or verbal 'assault'[6]. Visibly different individuals often exhibit problems involving spirals of negative emotions (for example, social anxiety), maladaptive thought processes (such as fear of negative social evaluation), unfavourable self-perceptions (for instance, low self-esteem and unfavourable body image), and negative behaviour patterns (such as excessive social avoidance)[7]. People born with craniofacial disfigurement report greater dissatisfaction with their facial appearance than their unaffected peers and experience discrimination in employment or social settings[8].

Contrary to the expectations of the lay public and many health care providers, research demonstrates that the extent, type, and severity of a disfigurement consistently fail to predict psychological distress[9,10]. A person's subjective perception of how noticeable their difference is to others is a better predictor of psychological and body image disturbance than is the assessment of a dispassionate observer or clinician[11]. Therefore, investigations focusing on individuals from their own perspective seem to be more informative.

Recent research has been focused on those attributes that appear to buffer a person against the stresses and strains of living with a visible difference. Some individuals appear able to cope well with being physically different, whereas others become socially disabled and psychologically distressed[12].

Research on how individuals with Crouzon syndrome manage their lives is lacking. Interventions for such individuals need to shift their interest from merely managing a pathological state to promoting adaptive coping strategies and benefit finding, aiming at improving well-being. The aim of this study, therefore, is to provide some insight on how young adults with Crouzon syndrome handle their life situation, focusing on their own perspective.

Material and methods

Study group and procedure

The study group consisted of six males and two females with Crouzon syndrome, with Caucasian ethnic background. The mean age was 25.4 years. They had a wide range of clinical presentations and reported that they had undergone many craniofacial surgeries, starting from their childhood years.

A letter about the study was sent out to 19 young adults with Crouzon syndrome (identified from the patient registry of the jaw orthopaedics unit of the Gothenburg university clinic of orthodontics) living all over Sweden, asking them whether they were willing to participate in the investigation. A reminder was sent a month later. If they replied positively, they were asked to contact one of the authors (U.H.) 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 min was conducted with each one of the participants in the study.

The study was approved by the Regional Research Ethics Committee of Gothenburg (Registration Number Ö 342-99).

Method

The qualitative research method of grounded theory was applied as described by Glaser and Strauss[13] and advanced by Strauss and Corbin[14], and later by Charmaz[15]. Grounded theory is an inductive research approach, which aims at generating concepts, models, or theories developed from empirical data, in order to explain and/or predict the phenomenon under study. Such methodology is usually employed in the exploration of rare and understudied social phenomena.

Data collection

Open audiotape-recorded interviews were conducted with the informants. The interviews started with the question 'how is it to live with Crouzon syndrome' and were held in a conversational style. 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 concerning themes such as *daily life*, *school situation*, *thoughts about meeting a partner*, *friends*, *and thoughts about the future*. Each interview acted as a guide for the next one. Nevertheless, as the possible participants of the study were few ($n = 8$), the reached saturation was judged to be tentative.

Data analysis

The audiotaped interviews were transcribed into text and through a line-by-line reading process, they were analysed by specific coding strategies[14]. These strategies included *open coding*, assignment of codes to the text based on words or phrases that captured meaning in the data; *axial coding*, comparing open codes with each other to create relevant more abstract categories; and *selective coding*, using frequently occurring codes to create a core category, which 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'.

Results

In the analysis of data, it became obvious that the participants' main concern, which was the core category, was *to make the best of their situation*. To be able to do so, they developed different strategies since childhood, in order to handle their vulnerable and exposed life situation and enhance their well-being. These strategies are presented below in five descriptive categories, labelled: *committed to an engaging activity*, *avoiding exposed situations*, *actively launching oneself*, *struggling with normalizing facial appearance*, and *lowering the expectations of finding a love partner* (Fig. 1). The more these strategies were used by the informants, the higher the possibility was that they could cope with their life situation and also experience increased well-being.

Short, exemplary quotations from the interviews, translated from Swedish, are presented below the strategies used, in order to illustrate these.

Committed to an engaging activity

Informants described how they had, already as children, found an engaging and time-consuming interest that they were devoted to, such as horseback riding, reading literature, or engagement in the church. The activities were of such kind that their facial appearance did not matter and were something they had scheduled to do, regardless of how they felt that day. After being involved in such a special interest, they felt less focused on their facial appearance during the performance of the activity and they felt an increased well-being.

… horseback riding … it became pretty interesting when I started to think a little more about it, because there I felt that I actually… it was probably one of those times when it wasn't so very important to have … I mean to be the perfect … that it wouldn't show or so …, but rather I could walk around there and be a little visually impaired, have a little Crouzon … the important thing was that I could handle my horse and riding, that I … there were other things that were important.

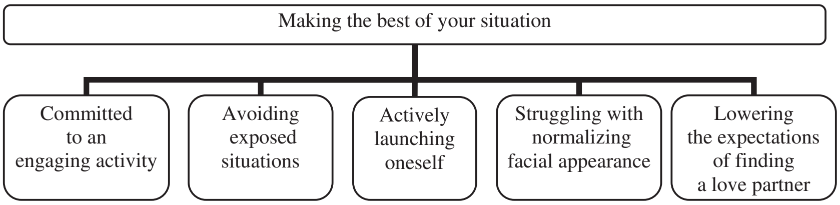

Fig. 1. A grounded theory model, depicting the relationship between the core category of concern for the Crouzon syndrome young adults interviewed and the relevant descriptive categories.

Avoiding exposed situations

The interviewed individuals with Crouzon syndrome described how they tried to avoid exposed situations, implying avoiding places where other people or strangers could stare at them. This made them feel like public persons and took away the possibility of them strolling anonymously in the streets and stop thinking about their life situation for a while. To be stared at and be pointed at also made them feel as outsiders and less worthy than others.

… but I don't feel normal at all since I am stared at and very often, you know … so I try to avoid ending up, for example, to end up in a tight spot… for example on a train or a bus you know, I used to board a railway car that didn't have that many people, you know …

Actively launching oneself

The participants in the study actively launched themselves at others they wanted to be acquainted with, or wanted to be friends with. When they met new persons or friends they struggled to be charismatic: open, social, humoristic, and focused on others, in order to help others to see behind their facial appearance. They also described how they felt suspicious towards others, and how others reacted to and thought of their facial appearance. Therefore, they carefully selected and rarely met new friends.

You can assume that you … I mean that … at the risk of sounding cold, it is actually so that people that don't look good, they have to have something else, you have to use your personality in a different way, but it is difficult to answer. I have a very … I have a very, as I see it, good sense of humour, laugh a lot at all sorts of funny things or what you would say … I have a very peculiar personality in that way …, but it is difficult, I think it is unavoidable that your personality is affected by what you have been through, I am quite convinced of that.

Struggling with normalizing facial appearance

Especially during their teenage periods, the interviewed individuals with Crouzon syndrome struggled with different kinds of surgical reconstructions, in order to normalize their facial appearance as much as possible. Some of these operations were especially physically and psychologically difficult and led to major facial changes, which in turn, led to identity crises. Furthermore, the informants had to spend significant time at the hospital during these surgical procedures, which also exposed them to other schoolmates as different. When they finally returned to school, it was with a changed facial appearance. During these facial and identity changes from major surgical interventions, the participants in the study thought that it was crucial to receive appropriate psychological support from both their families and friends, as well as from professionals. The more the facial appearance was normalized through surgical operations, the less the informants felt that they suffered from a syndrome and the better they could handle their life situation, thus experiencing enhanced well-being.

And even this waking up with a new face after the operation, it was also a pretty big shock in itself, so it was a change, it was like you felt that you didn't recognize yourself … That in some way … you got a new face and you were like, you know simply unsure of who you were, and that you simply didn't recognize yourself appearance wise. That was a pretty big shock right there…

Lowering the expectations of finding a love partner

The informants had, already as teenagers, a wish for meeting a love partner that would love them just the way they were, someone who could see beyond their facial appearance. However, at the same time, they argued that it would be probably difficult. If the individuals with Crouzon syndrome tried to find a love partner, they lowered their choice of partner accordingly. In effect, they preferred to fall in love or long for somebody that also had an exposed life situation and became vulnerable. During their teenage period, when their schoolmates and friends found different love partners, the informants felt locked out from this part of life. This made them feel as outsiders that differed from the others. If the informants finally met a love partner, that significantly increased their potential to handle their life situation. It seemed in the interviews that the more the informants' facial appearance was normalized, the higher the possibility was to find a love partner.

Yes, of course, I had, but I realized that it would probably be difficult, difficult to meet someone, you know, that might like you … I never thought of it, I've always taken it for granted …

Discussion

In this study, young adults with Crouzon syndrome proved to be interested and able to discuss how their lives are affected due to their specific physical characteristics. The main finding of the present investigation, as depicted by the core category of our results '*to make the best of their situation*', shows that even in adverse conditions, as in Crouzon syndrome, several individuals do find ways to live with their difference and to succeed in various aspects of life, using methods they construct. This is consistent with the findings of Strauss and Fenson[16], who concluded that in people with craniofacial anomalies, in spite of numerous challenges, healing occurs and they join nonaffected persons in the search for meaning and quality in their lives. Learning to cope relates to the concept of resilience[17], which has been described as 'normal development under difficult conditions'[18], and is recently seen as a dynamic process involving an interaction between both risk and protective processes, internal and external to the individual, that act to modify the effects of an adverse life event[19]. Although a positive way of life may not be true for all of the young adults with Crouzon syndrome, the possibility that some of them can overcome the barriers that their syndrome places should be included in a balanced psychological assessment of their situation.

It seems that the commitment of individuals with Crouzon syndrome to engaging activities, where their facial appearance does not matter, can be regarded as an effective coping strategy to handle their condition. Diverting energy and attention from the facial difference to other meaningful aspects of life can improve well-being. Faith, religious belief, and inner spiritual strength have also been cited as significant sources of comfort and meaning[16].

Individual differences in the coping strategies were noted in our study. People employ a variety of emotion- and problem-focused coping strategies that are related to either the 'protection of the self'[20] or to 'the presentation of the self'[21]. In the present investigation, the former can be seen in the descriptive category of '*avoiding exposed situations*', whereas the latter manifests in the category '*actively launching oneself*'. Self-protection strategies include the use of social comparisons, and denial and external attributions. Self-presentation strategies refer to behavioural and cognitive strategies used to maintain a sense of acceptability or self-esteem against the impact of others' reactions.

Galvin[22] found, in his study of identity transformation following disability, that people who constructed positive identities following impairment were those who conceptualized their adversity not as a personal failure, but as a socially mediated phenomenon, that could be challenged and resisted. The role of society towards the ones who are visibly different seems to be highly significant.

In this study, '*struggling with normalizing facial appearance*' was a key descriptive category. There is no doubt that improving the facial appearance on patients with Crouzon syndrome with the help of surgical reconstructions can offer obvious benefits. However, these interventions are not a panacea. Rumsey and Harcourt[7] discuss that in subscribing to the biomedical view of a simple relationship between appearance and adjustment, care providers are colluding with the myth that quality of life necessarily improves when physical appearance is enhanced. For some people, one operation may suffice, but many undergo multiple surgeries in the quest for a 'normal' appearance. The question of 'when to stop' can be complex. Patients' and clinicians' understandings of 'improvement' and their expectations of treatment may differ fundamentally. The patients' own perspective, desires, and expectations should be given the highest attention. Research on cleft lip and palate patients has demonstrated that a significant proportion of adolescents feel left out of treatment decisions in the craniofacial setting and that, when questioned, their satisfaction with clinical outcomes often do not correlate with their parents', who may be making the treatment decisions for them[23]. If the same holds true for patients with Crouzon syndrome, it remains to be validated.

An important issue illustrated from this study is that following craniofacial reconstruction, the reactions to a surgically produced result may not be universally positive. Those accustomed to their difference may feel the altered appearance 'is not the real me', leading to identity crises. These crises can have an additive effect on teenage crises. During the teenage period of life, the self is not fully developed, leading to an increased vulnerability in changes of the appearance or the self. Therefore, psychological counselling is of great importance to individuals undergoing craniofacial surgical reconstructions including young children, teenagers, and adults.

Sarwer *et al.*[8], in their investigation of psychological functioning of individuals born with craniofacial anomalies, found that more than 70% of such individuals indicated that others had not wanted to become romantically involved with them, because of their appearance. This is in line with our results as described in the category '*lowering the expectations of finding a love partner*'.

Living with Crouzon syndrome concerns a social process that has been sparsely described to date. Most relevant studies are heterogeneous, 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. The present investigation qualitatively studies a well-defined group of individuals, where all have the same diagnosis of Crouzon syndrome.

A grounded theory approach is particularly suitable in the investigation of social processes[13]. It has an enhanced ability to generate fresh theoretical insights directly from the data, helping the participants to tell their stories in ways that are not influenced by any preconceived ideas on the part of the researcher. Therefore, grounded theory was considered an appropriate research method for the present investigation.

The limitations of the current study should be taken into account. First, the preference to interview over the telephone was at the expense of missing some aspects of nonverbal communication with the participants of the study. Nevertheless, this had the advantage of reducing possible inferiority feelings of the informants during the interviews. Secondly, the individuals with Crouzon syndrome that participated in the current investigation may not represent the whole population with Crouzon syndrome in Sweden. The syndrome shows variable clinical severity and psychosocial adjustment; therefore, less outgoing individuals with Crouzon syndrome may experience different clinical and social situations. Finally, due to the low prevalence of the syndrome, the participants in this investigation were few. Therefore, the results presented are referred to as tentative results, although a saturation of information was believed to have been accomplished. Nevertheless, according to Charmaz[15], the unit of analysis in a grounded theory study concerns *events and stories*, rather than the *participants per se*. Thus, the number of informants is less interesting than the content and the quality of the data.

The findings of our study are applicable to young adults with Crouzon syndrome living in Sweden, as their validity refers to individuals living under the same cultural and natural context as the participants. Furthermore, it is wise to avoid generalizations of results from a single qualitative study. Rather, the presented results should be interpreted as a substantive model for understanding how young adults with Crouzon syndrome experience their lives. In other words, the present study (as the case is with every qualitative study) should be viewed as a hypothesis generating study, waiting for confirmation from future quantitative studies.

In conclusion, the present investigation indicates that the main concern of the participants in our study was *'to make the best of their situation'*, which depicts an attitude towards life to handle their condition. The adaptation of successful coping strategies seemed to be crucial in the quest of attainment of higher self-esteem and of more positive self-image. The more the persons with Crouzon syndrome had developed the described coping strategies over time, the more they became able to handle their life situation. This skill led to enhanced well-being.

What this paper adds

- Although young adults with Crouzon syndrome suffer from obvious facial dysmorphic features due to the condition, this study acknowledges that some of them may find ways to handle their vulnerable life situation and enhance their well-being.

Why this paper is important to paediatric dentists

- Physical appearance concerns are at a peak during adolescence and young adulthood; therefore, patients with Crouzon syndrome may have tough experiences at this particular time.
- The concept of patient-centred care involves understanding patients' views and concerns with respect to their clinical condition and its effect to their everyday life. Acknowledging the specific psychosocial issues affecting these patients is likely to enhance patient satisfaction with the care received.

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