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PERCEPTION OF CHILDREN WITH CRANIOFACIAL MICROSOMIA ABNORMALITIES BY THEIR CLOSE RELATIVES*

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SUMMARY

Background: When a child is born with a deformed face, his/her social environment is quickly confronted with a change which undoubtedly favours numerous over-generalizations. Children with craniofacial microsomia anomalies (CFCs) may experience less social support, more rejection and make them withdraw from their social life more frequently. The purpose of this research study is to show how children with craniofacial hypoplasia abnormalities is perceived by their close relatives.

Material/ Methods: The sample of this study consisted of 26 participants (F=16; M=10). This research study was conducted using the following questionnaire methods: the Scale of Over-Generalization Effect by K. Milska and A. Mański (*SOGE*), modified Own Health Assessment Scale method (*SOWC*) and the Authors-Designed Questionnaire (*ADQ*) to obtain information on the issue.

Results: A child's illness always affects a variety of family life spheres. Most of the surveyed women did not experience any complication during pregnancy or childbirth. Unfortunately, after their child was diagnosed, most adults were not offered getting in contact with a psychologist / psychiatrist / psychotherapist. Some relatives – after the birth of a child with craniofacial microsomia (CFM) – reduced their working hours or gave up work completely in order to take care of the child. The most urgent needs for this child's illness reported by adults most often referred to educational and financial matters. At the time of this research study, most of the respondents (61%) – upon the birth of a child with CFM – considered it plausible to enlarge their family.

Conclusions: As a result of the conducted research studies, new variable systems (a type and character of dysmorphia, closeness – distance) were identified, which may be a relevant element to facilitate research studies on the perception of children with body deformities by their environment. In the characteristics of a child with CFM, it was shown that his/her close relatives evaluate the child positively. Family members apply constructive strategies for coping with the child's illness, however, the research study results indicate the legitimacy of introduction of psychological and psychiatric consultations for close relatives to the standards of CFM child treatment.

Key words: neurodevelopmental disorders, craniofacial dysmorphy, overgeneralization, anomalous face overgeneralization, microgenetic theory

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BACKGROUND

Craniofacial microsomia (*CFM*) is a complex, congenital disorder which affects person's key facial parts. In the literature on the subject, CFM is characterised to be a spectrum of anomalies starting from isolated, unilateral facial defects within selected face parts up to bilateral malformations of ears, mandible, soft facial tissues and eye surroundings [1]. Under CFM the following two complex syndromes are most often diagnosed: hemifacial microsomia (*HFM*) and oculo-auriculo-vertebral dysplasia (*OAVS*). If facial abnormalities are found on the left or right side, HFM is the most common diagnosis. In the case of OAVS, the diagnostic process is more complicated because changes which form the so-called classical triad of changes should be singled out: mandibular underdevelopment resulting in facial asymmetry, eye and ear deformations and spine changes [2,3]. It is worth mentioning that – apart from the structural changes mentioned above – the following abnormalities may often occur in the CFM spectrum: upper respiratory tract obstruction, dysphagia, speech and hearing disorders and central nervous system defects [1]. As numerous authors point out [4,5,6,7], it is difficult to unequivocally explain the origin of neurodevelopmental disorders which occur under the CFM spectrum. When intellectual disability occurs, dominantly it is at mild and moderate level, i.e. children with intellectual disabilities experience learning difficulties and emotional disorders relatively often and they are only occasionally diagnosed with severe intellectual disabilities. As Guralnick [6,7] points out, negative behavioural effects can be effectively smoothened by accurate and reliable functional diagnosis and early implementation of rehabilitation and therapeutic effects. Riklin et al.[8] made a review of more than 35 articles on adolescents with CFM and their psychosocial behaviour and indicated that low social skills and negative interactions with peers are an inherent aspect of such disorders. The coexistence of these features in the light of the data provided may result in, among others, a lack of self-confidence, loneliness, little positive support and also much self-discontent. The authors of this paper also indicate in their theoretical analyses that a face itself and its adaptive role in interactions with other people may play an important role in the emergence of difficulties in social relations kept by CFM-affected people.

From a psychological perspective, a person's face is an extraordinary source of impressions which let us to acquire knowledge about other people. The acquisition of this knowledge undoubtedly takes place in interactions between and among people. Relatively recently, a number of factors related to the physicality of a given person's face have been discovered and described which very strongly determine his or her 'picture' in other people'sminds. Zebrowitz and Montepare [9] defined this method of deduction as *overgeneralization*. Unusually often this type of deduction is related to the physical attractiveness of an individual person's face (*anomalous face overgeneralization*). Griffin and Langlois [10] point out that people tend to perceive persons with unattractive faces in a more negative manner. As a number of the authors emphasise [11,12,13], this manner of attribution

of certain features related to interpersonal space has a adaptative value. Quite often such a changed face is interpreted as powerlessness or impuissance of its owner, presence of certain diseases or gene abnormalities [9]. It is worth noting that the term "dysmorphia" stands for an extremely broad spectrum of morphological anomalies to be perceived on one's face [14]. Dysmorphic changes are common in clinical practice and may include:

- a lack of certain anatomical parts on the face;
- changes in the structure of certain facial parts consisting in breaking its continuity within one or more areas;
- disorders in the development of a given part (e.g. a malformed auricle, split nose or wide mouth, etc.);
- displacements of selected facial and masticatory organ elements, e.g. Pierre Robin sequence, narrowly spaced eyes in Cri Du Chat syndrome, slanted palpebral fissures in Down syndrome;
- changes in the profile, features and proportions of facial parts (e.g. as a holoprosencephaly effect, Edwards syndrome, Patau syndrome, etc.) [15].

It is worth pointing out that facial dysmorphia is congenital. A face gets developed between 4 and 8 weeks of foetal life, and at the end of the embryonic period it takes on a completely human appearance. When a child is born with a deformed face, his/her social environment is quickly confronted with a change which undoubtedly favours numerous over-generalizations. As mentioned earlier [8], adolescents with craniofacial microsomia anomalies (CFCs) may experience less social support, more rejection and make them withdraw from their social life more frequently. This kind of experience many a time build grounds for CFCs patients' future social interactions. It is easy to notice that their existing difficulties in social relations may have a secondary nature and over-generalizations of healthy people who "happen to took part" in relations to people with CFCs may be their primary source. In turn, the research studies made by Mański [16,17] show that facial dysmorphia suffered by children with moderate intellectual disabilities may remain unnoticed by their mothers and educators in special education centres. In the context of these studies, relevant dependencies were detected between the image of a child and his/her personality, disease pattern and mother's and teachers' value system. The detected phenomenon made scientists and practitioners focus on interpersonal factors related to people involved in diagnostics, treatment and rehabilitation of people with facial deformities. How they perceive a person with facial dysmorphia not only depends on a level of their professional competence but also on numerous personal specificities, including the image of bonds (*the emotional dimension of their relations*) built in the process of diagnostics, treatment and broadly-understood rehabilitation [18,16].

MATERIAL

The research study was performed at the conference organised for carers of children with craniofacial malformations, which took place at the Facial Cranio-

facial Surgery Centre with Department of Maxillofacial and Reconstructive Surgery in Olsztyn and during individual meetings with carers while their visits to the Psychological Counselling Centre for Rare Genetic Diseases at the Institute of Psychology of the University of Gdańsk.

The research study involved 26 legal guardians of CFM-affected children, 14 of them were married (7 couples). The other participants formed a group consisting of one parent of each child (*n*=11) and one person with the legal status of a minor person's guardian who is not his/her biological parent (grandmother) (*n*=1). The study group consisted of 16 women and 10 men aged from 30 to 52 years (*M*=38.1 years; *SD*=6.2). 90% of the respondents were married, the rest were singles or divorced persons.An information note: the limitation connected with number of participants (close relatives) is due to the CFM which is one of the rare diseases spectrum. The participation in the study was voluntary, the study was made after the consent of the participants. This scientific project was approved by the Independent Bioethical Committee for Scientific Research at the Medical University of Gdańsk (NKBBN/48/2017) and the management of the Provincial Specialist Children's Hospital named after Prof. Stanisław Popowski in Olsztyn.

METHODS

The research study was conducted with the application of questionnaire methods. The following research methods were applied: the Authors-Designed Questionnaire (*ADQ*), the Scale of Over-Generalization Effect (*SEN*) by K. Milska and A. Mański and the modified Own Disease Assessment Scale (*SOWC*) by S. Steuden. Below is a brief description of the applied research methods.

The Authors-Designed Questionnaire (*ADQ*) used in the study was intended to obtain basic information about the respondents. It covered a set of questions referred to such exemplary variables as: gender, education, place of living, etc. Additionally, the respondents were asked about circumstances of getting a CFM diagnosis and influence of this disease on the social and family standing of child's relatives.

The Scale of Over-Generalization Effectcontains 20 pairs of adjectives with opposite meaning. Each pair forms a bipolar 1-7-point scale which lets to evaluate a particular feature. Based on the constructed profile showing a level / intensity of the features specified in the *SEN* method, the following information can be obtained: a general picture of an individual child, distribution of four factors and their intensity (*Dexterity, Personality traits, Value and Appearance*) as well as a profile of 20 individual characteristics [16].

The Own Disease Assessment Scale by S. Steuden was modified as required for this study. A change was made to the reference point in the assessment of each item. The respondents were asked to assess the relevance of their child's illness (instead of the originally determined relevance of their own illness). The respondents' task was to express their opinion to each statement at the 5-point Likert scale. Here is an example of such a statement: "*For me, my child's illness*

is a loss of hope to accomplish my personal life plans." The provided method consists of 6 thematic scales and 1 control scale. Among the thematic scales, 3 of them (*Challenge, Value* and *Benefit*) refer to the illness in a positive manner and 3 ones (*Threat, Obstacle / Loss, Harm*) let to make a statement on its negative aspects under the psychological pattern of this disease. The control scale (*Relevance*) is the starting point for all kinds of analyses because it shows a level of balance between health and illness, indicating the potential dominance of any of these two conditions [19].

RESULTS

Perinatal context

In most families (58%) no complications related to the child or mother's health during pregnancy were reported. Nearly 79% of women did not experience any difficulties during childbirth. More than half of the surveyed carers (54.5%) got to know about the child's disease at the moment of his/her birth, 27% of them heard the diagnosis during the first year of their child's life, and 14% – at later stages of his/her development. There was only one person (4.5%) who learnt about the child's developmental abnormalities during the prenatal period. In spite of potential difficulties related to the birth of a disabled child, 83% of the respondents were not offered to get in touch with a mental health specialist. Only 17% of them obtained information on such support and 13% – made use of this option. Further in time, due to the child's illness and related circumstances, 33% of the respondents started visiting a psychologist, psychiatrist or psychotherapist.

A child with CFM was born first (as the first child) in 44% of the surveyed families, second in 44% of these families and third in 12% of them. As many as 79%

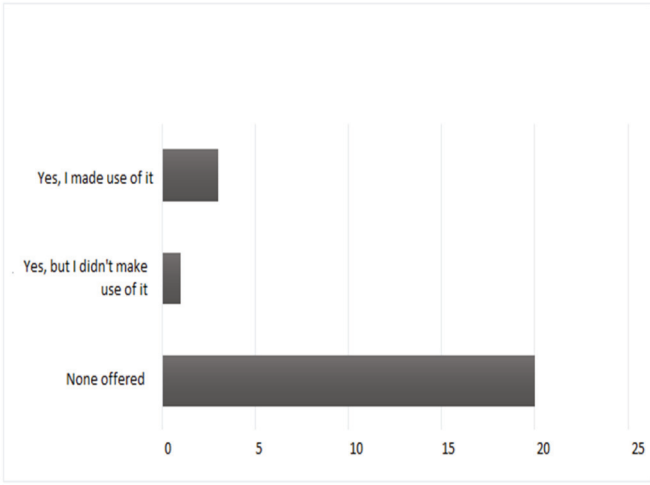

Fig. 1. Number of CFM-affected child's legal guardians in the context of offered and obtained specialist assistance (psychiatrist / psychologist / psychotherapist)

of the surveyed families – after the birth of their child with facial dysmorphia – had no more children, and 21% of such families had more. During the study period, 39% of the surveyed carers declared no further reproductive plans.

Education and professional activity of the respondents

More than half (58%) of the surveyed carers had higher education. The second place – in terms of numbers – was occupied by the respondents with secondary education (34%), while the least numerous group was composed of people with vocational education (8%). Nobody in the study group declared primary education.

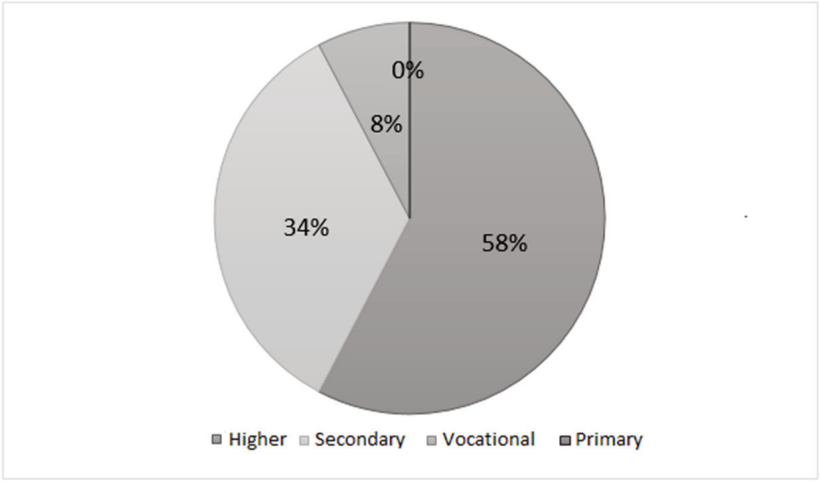

Fig. 2. Level of education of the surveyed carers

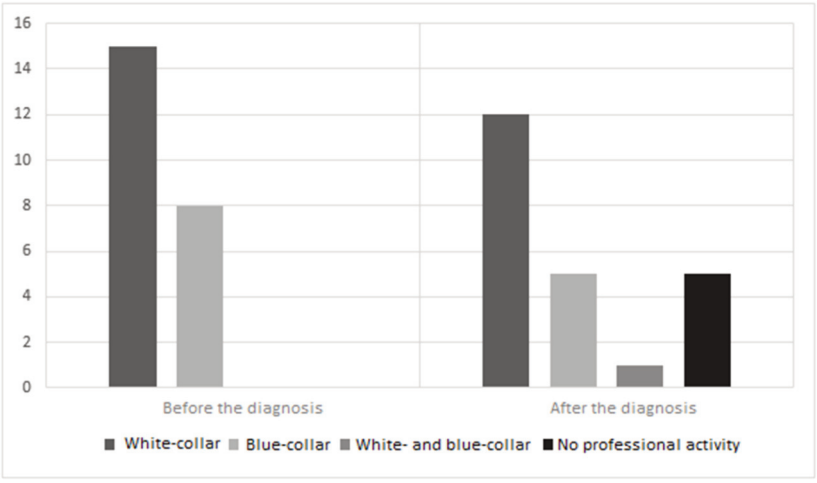

Fig. 3. Type of professional activity of the surveyed carers performed before and after child's diagnosis

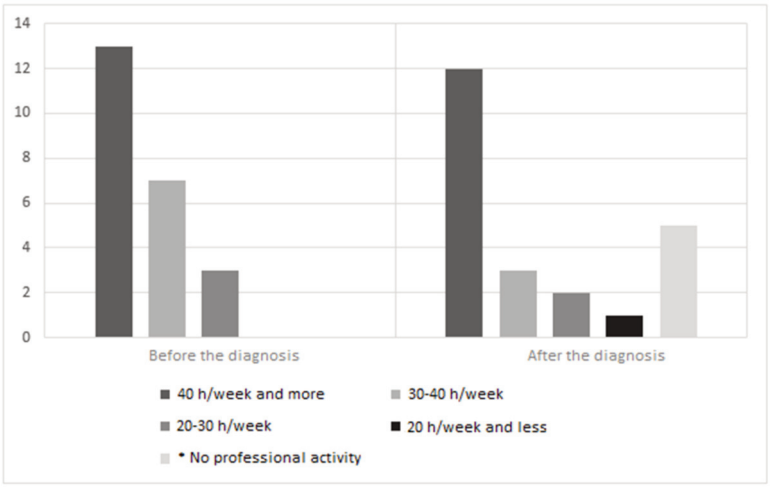

Fig. 4. Working hours of the surveyed carers before and after the child's diagnosis

Before child's diagnosis was made (among the respondents – most often until the child finished the first year of life), 15 of the respondents performed white-collar work, while 8 of them – blue-collar work. After child's diagnosis is made, a number of people performing both white-collar and blue-collar work decreased (*n*=12 and *n*=5, respectively), while a number of the respondents who did not take up professional activity increased (*n*=5). One surveyed person declared to perform combined white- and blue-collar work.

Taking into account such changes in the range of work performed among the surveyed group, time spent on professional activity undertaken by the respondents was analysed.

Before child's diagnosis, in the surveyed group 13 persons (57%) worked full time or more, 7 persons (30%) spent between 30 and 40 hours a week at work, while the working time of 3 persons (13%) was between 20 and 30 hours a week. After the disease was diagnosed, a number of the respondents who worked 30 hours or more a week (65%) decreased. 2 respondents (9%) took up a 20-30 hour/week job, 1 person (4.5%) – maximum part-time, while as many as 5 people (22%) gave up professional activity.

Needs arising from the child's illness

More than half of the respondents (*n*=15) considered the child's educational needs and requirements related to their financial standing to be the most relevant needs arising from the child's illness (*n*=13). 7 people pointed out educational needs of their child's siblings. A slightly smaller number of the carers declared other needs (*n*=6) or admitted that they needed rehabilitation equipment (*n*=5). One respondent pointed out needs related to the child's flat furnishing.

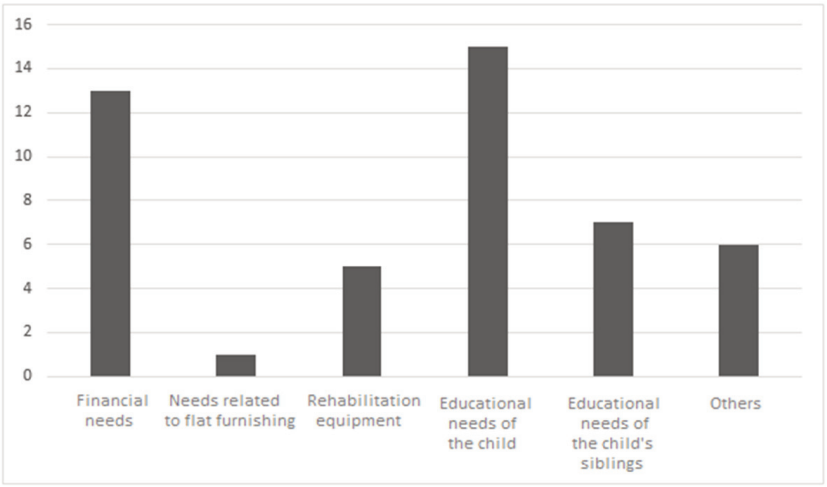

Fig. 5. Profile of needs arising from the child's illness from the perspective of the surveyed carers

Perception of CFM-affected children by their closest persons

The mean result of the overall picture of children with CFM obtained by their closest relatives was equal to *M*=5.38. The conducted analysis lets to conclude that, apart from *Value* and *Appearance*, all the mean results of the examined factors are above average (*Dexterity* and *Personality traits*). The caregivers of children with CFM scored the highest in terms of *Dexterity* (*M*=5.70); a slightly lower score was obtained when assessing the child's *Personality traits* (*M*=5.41). The analysis of the overall result and the mean results of the examined factors lets to conclude that positive pictures of children with CFM stay in the minds of their closest persons.

An in-depth analysis of the mean results obtained by close relatives in each scale indicates a variety of images of a child with CFM, with a predominance of positive assessments. Close relatives assessed their child highly positively in the following scales: Scale I (*M*=6.44), Scale II (*M*=6.61), Scale IV (M=6.22), Scale IX (*M*=6.33), Scale XI (*M*=6.39) and Scale XVII (*M*=6.17). Positive assessments were obtained in the group of close relatives of CFM-affected children in the following scales: Scale III (*M*=4.50), Scale V (*M*=4.78), Scale VI (*M*=4.78), Scale VII (*M*=4.61), Scale VIII (*M*=5.78), Scale X (*M*=4.50), Scale XII (*M*=5.56), Scale

Table 1. Mean (M) and standard deviation (SD) for the factors and the overall result based on the feedback provided by the carers of CFM-affected children

The Scale of Over-Generalization Effect (*SEN*)	Carers of CFM-affected children	
	M	*SD*
Factor: *Dexterity*	5.70	2.06
Factor: *Personality traits*	5.41	1.97
Factor: *Value*	5.22	2.04
Factor: *Appearance*	5.20	2.05
Factor: *Overall result*	5.38	2.03

Table 2. Mean (M) and standard deviation (SD) for the results of individual DS scales at the surveyed CFM-affected children's close relatives

SEN	*N*=18	
	M	*SD*
I. Active – Passive	6.44	1.15
II. Emotional - Non-emotional	6.61	0.61
III. Precise – **I**mprecise	4.50	2.13
IV. Slim – Obese	6.22	1.11
V. Tough - Prone to getting tired	4.78	2.29
V**I.** Empathic - Non-empathic	4.78	2.12
V**II.** Responsible – **I**rresponsible	4.61	2.19
V**III.** Pretty – Ugly	5.78	2.18
IX. Apt – **I**napt	6.33	1.97
X. Reflective – **I**mpulsive	4.50	2.13
X**I.** Rational - Non-rational	6.39	1.04
X**II.** Motionally fit - Motionally clumsy	5.56	1.85
X**III.** Self-reliant - Non-self-reliant	4.78	2.46
X**I**V. Courageous - Lacking courage	5.39	1.88
XV. Sacrificial - Non-sacrificial	5.67	1.89
XV**I.** High – Low	5.06	2.15
XV**II.** Physically strong - Physically weak	6.17	1.30
XV**III.** Social - Non-social	5.78	1.93
X**I**X. Hard-working – Lazy	4.94	2.11
XX. Not-deformed – Deformed	3.39	1.61

XIII (*M*=4.78), Scale XIV (*M*=5.39), Scale XV (*M*=5.67), Scale XVI (*M*=5.06), Scale XVIII (*M*=5.78) and Scale XIX (*M*=4.94). The surveyed persons obtained the lowest mean score by assessing their children in the Scale XX (*M* =3.39). Close relatives perceive their CFM-affected children as more active, more apt and physically stronger. These features are part of the *Dexterity*factor. Moreover, close relatives indicate that their children are more emotional and less obese. Only one trait related to a level of deformity of the child's body with CFM was assessed negatively by relatives.

Perception of the child's illness with CFM by close relatives

Close relatives perceive their child's CFM illness as a phenomenon of medium relevance in their lives, which is reflected in the *Relevance* Scale (*M*=2.90). In the daily life of the surveyed relatives, their child's illness is a challenge (*M*=3.75)

Table 3. Mean (M) and standard deviation (SD) for the results in the Own Disease Assessment Scale (the modified version) at the surveyed CFM-affected children's close relatives

Own Health Assessment Scale (*SOWC*) (modified version)	*N*=23	
	M	SD
THREAT	2.54	0.91
BENEFIT	1.92	0.49
OBSTACLE	1.90	0.81
CHALLENGE	3.75	0.82
HARM	2.04	1.02
VALUE	3.03	1.09
RELEVANCE	2.90	0.66

and experience with its special value (*M*=3.03) to them. It is worth mentioning that the singled-out parameters are positive aspects in the psychological picture of this disease. The analysis of the results of the other parameters shows that the psychological picture of this disease is not dominated by negative aspects (*Obstacle, Harm, Threat*). Their close relatives are not inclined to define this illness as an incident or event which gives rise to certain privileges (Benefit).

DISCUSSION

The birth of a child with a disability is a turning point in the life of an individual family. It poses a challenge to the closest relatives of ill children through the reorganisation of their existing life. Such changes can be multidimensional, concerning the family itself, professional standing of its members, as well as far-reaching future plans. It seems that this kind of changes may affect women more, because it is they who most often provide care for the sick child [20].

The birth of a child with a disability severely determining his/her appearance is an event which triggers a number of changes, especially in the psychological sphere of his/her close relatives. The behaviour and operation of relatives in this case can be explained – on one hand – on the grounds of the model of adaptation to this disease / disability [21] and – on the other hand – in the context of the concept of psychological mourning [22]. The results of the Author's Survey showed that the adaptation process had a significant impact on: close relatives' procreation plans, changes in the character of their professional activity and working time after the child is diagnosed with CFM. Resignation from having children after the birth of a child with CFM (71%) can be explained at least in two ways. The first one may be related to their fear of having more child with disabilities. The second one refers to overall reflections on the very broad range of interactions which carers need to take to help their child with CFM. In this context, there is not enough space to take care of one more child. Further changes concern the area of professional activity. Some respondents (20%) did not take up their professional activity after their child was diagnosed with CF and a similar number (22%) decided to change working hours and character of activities in their working environment. These relevant adaptive changes beyond the family environment (but supporting it) testify to the existing awareness that children with CFM should be appropriately treated.

Both the model of adaptation to the disease / disability [21] and the concept of mourning by M. Bristor [22] present the phase-by-phase process of adaptation. Both of them start with the shock phase, which tends to be extremely unstable and full with strong negative emotions. The vast majority of the respondents (83%) were not offered any contact with a mental health specialist shortly after their child with CFM was born. It is not easy to explain this state of affairs. On one hand, it is true that under the CFM treatment standards, the priority from the very beginning is to keep a child with CFM alive, and then - on the other hand - the need for reconstruction of facial elements is stressed. On the other hand,

close relatives when in shock may find it very difficult to express any demands or needs related to their child.

When a disability affects facial areas, it can affect the 'quality' of child's interactions with his/her environment. As the face plays an important role in the process of receiving and transmitting messages, people around may feel it difficult to acquire and develop their knowledge about such a child with disabilities. As Brzezińska et al. notices, "*the key issue is whether parents and caregivers are aware that the child is in the foreground, or whether it is the disease / disability*" [23]. In the perception of a child with a disability, the author postulates to make a distinction between a domain related to the disease / disability (the perception of disease) and a domain related to the child (the perception of child's healthy areas). A similar suggestion was made by Kościelska [24] who pointed out that in case of children with disabilities it is necessary to take into consideration the so-called areas of normality.

In relation to CFM-affected child's close relatives, the above-specified ranges of perception were estimated. In terms of the child image, high and medium positive characteristics were obtained, except for the one describing the subjective level of CFM-affected child deformity (M=3.39). The provided results contradict the Zebrowitz and Montepare theory [9] because the most frequent reaction to facial deformities is to depreciate features lying beyond the appearance of a disabled person (e.g. personal traits, dexterity, values, etc.). Moreover, the low assessment of a child with CFM in terms of the non-deformed – deformed feature differs from the results obtained in the studies by Mański [16,17], where the author discovered that mothers and teachers of children with moderate intellectual disabilities and facial dysmorphias did not notice such dysmorphias (high assessment in the scale XX). However, it referred to children who had numerous minor dysmorphias which made their unique facial appearance (e.g. Down syndrome, Williams syndrome, etc.). Therefore, it can be concluded that the nature and type of anomalies on the face can also play a relevant role in the attribution of certain characteristics to a child with disabilities. Many minor dysmorphias are difficult to define by non-specialists. Their size and number and often lack of effects on the key vital functions (e.g. breathing, swallowing, seeing and hearing) are hardly grasped by close relatives. Unlike the identification of CFM defects. From the very beginning they can be noticed and their presence significantly affects the basic child's functions.

Turning to the picture of a child's illness / disability it is worth noting that there is a specific balance between the perception of healthy and sick child's areas (*Relevance* – 2.90) in the consciousness of CFM-affected child's close relatives. Looking from the biopsychosocial perspective, it may mean that relatives tend to fulfil their child's needs in a harmonious manner. They will not tend to omit any of the key areas specified in this concept. The factor 'Relevance' outlines the manner of further analysis of positive and negative areas in the psychological picture of this illness. However, its positive aspects dominate in the study group (*Challenge* and *Value*). The perception of this disease as a challenge indicates that close relatives can go on in the phase of their constructive adaptation to the

child's illness (the adaptation model). The higher results of this parameter are also a sensitive marker of the psychological resources of surrounding people and the presence of people who are factually willing to help. Against the background of this group, the increased results under the *Value* can be interpreted in a completely different manner. As Steuden et al. [19] mentions, this factor shows that this kind of illness can become an incident or event with a deeper relevance not only for the affected person. Paradoxically, this experience becomes a driving force of self-fulfilment which lets not only to actively counteract its effects, but also to develop people from within one's immediate environment. In the context of deliberations made by Brzezińska et al. [23], the perception of a child with CFM by people close to him/her – assuming that the disease / disability does not obscure the whole picture of this child – was captured. This condition induces more to activate positive energy to tackle this child's disease rather than negative one causing adaptation difficulties. The obtained results correspond well with the studies by Mański [16,17], in which the distribution of parameters and their intensity in the psychological picture of this disease was alike, apart from the Relevance [3]. The author studies the perception of children with intellectual disabilities and dysmorphic facial abnormalities by mothers of these children. It is worth mentioning that too high results obtained at the primary level (Relevance) may adversely affect the adaptation of relatives towards the child's disease / child disability. This level may indicate that the illness is too much of a disturbance and requires finding new layers of energy and vitality. Finally, it seems necessary to provide potential explanations for the results obtained. Where do such discrepancies come from when confronted with the Zebrowitz & Montepare concept and the research studies by Mański?

It seems that emotional closeness to the child may play the key role here. A level of 'knowledge' of the child and the nature of bonds with him/her may influence the effects of over-generalization. Individuals have insight into their child at levels which cannot be reached by others (strangers). As for the CFM-affected child's close relatives, even more objective assessment on the degree of child's face deformity did not prevent them from giving their child high scores in terms of dexterity, personality traits and value.

The results might be interpret by microgenetic theory. An unpleasant real experiences (eg. the perception of deformities) releases negative emotions (e.g. fear, sadness) because it stimulates the lateral prefrontal cortex and strengthens the penalty system by experiences felt. Negative emotions e, are included in the working memory, and, they are remembered in the long term memory, if they are important for close relatives. At the same time, the reward system is weakened (cf. Fig. 4). Accordingly, pleasant dreams or experiences release positive emotions (e.g. joy), because they stimulate the reward system by creating connections from the basal part of the frontal cortex to the anterior (emotional) part of the anterior cingulate cortex. At the same time, the penalty system is weakened. The strength and duration of these emotions are associated with the importance of the type of malformations (minor dysmorphias / major malformations).

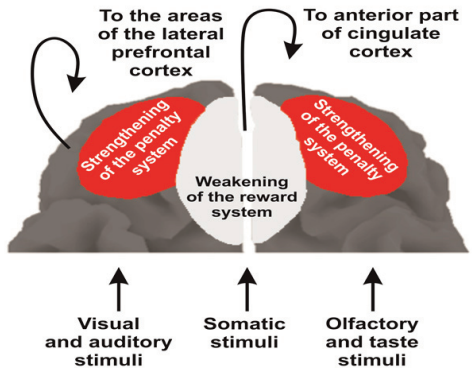

Fig. 6. The panelty and reward system
Source: M. Pąchalska [25]

Therefore, the type of malformations might modify the minimal (working) self, and the longitudinal (autobiographical) self, strengthening the significance of a given (negative or positive) event.

However, it should be stressed, that closeness may prove to be a valuable source of weakening the perception of a CMF child as a less attractive partner in interpersonal relations. When the perception of deformities by close relatives is concerned, a lack of compliance with the research studies Mański [16] directly results from the type of malformations (numerous minor dysmorphias / major malformations).

Based on the conducted research studies and confrontation with the results of the research by Mański [16,17], there is an emerging need to emphasize this variable in further research studies.

CONCLUSIONS

1. In the light of the provided results, it seems necessary to introduce psychological and psychiatric consultations for close relatives into the standards of treatment of children with CFM.
2. In the characteristics of CFM-affected children, close relatives provide positive assessments on them. Particularly high scores were recorded in terms of dexterity, aptness and physical strength. This pattern of traits may reflect the actual state or priorities which express their dreams and wishes related to the child.
3. The results obtained in confrontation with the concept developed by Zebrowitz and Montepare as well as research studies by Mański let to distinguish new systems of variables (a type and character of dysmorphia, closeness – distance) which may deepen and enrich the course of research studies on the perception of a child with body deformities by surrounding people.

4. People close to a child with CFM perceive his/her disease as a challenge and value, with its medium relevance for their daily operation. This system of parameters shows that very constructive strategies for dealing with the illness are applied by the child's close relatives.

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REFERENCES

Heike, C.L., Luquetti, D.V., Hing, A.V. (2014). Craniofacial microsomia overview. IN: M.P. Adam, H.H. Ardinger, R.A. Pagon, S.E. Wallace, L.J.H. Bean, K. Stephens, A. Amemiya, editors (ed.), *GeneReviews((R))*[Internet] (pp. 1-32). Seattle (WA): University of Washington.

Goswami, M., Bhushan, U., Jangra, B. (2016). Goldenhar Syndrome: A Case Report with Review. *International Journal of Clinical Pediatric Dentistry, 9*(3), 278–280. DOI: http://doi.org/10.5005/jp-journals-10005-1377.

Berker, N., Acaroğlu, G., Soykan, E. (2004). Goldenhar's syndrome (oculo-auriculo-vertebral dysplasia) with congenital facial nerve palsy. *Yonsei Medical Journal, 45*(1), 157–160. DOI: 10.3349/ymj.2004.45.1.157.

Padwa, B. L., Evans, C. A., Pillemer, F. (1991). Psychosocial adjustment in children with hemifacial microsomia and other craniofacial deformities. *Cleft Palate-Craniofacial Journal, 28*(4), 354–359.

Pillemer, F., Cook, K. V. (1989). The psychosocial adjustment of pediatric craniofacial patients after surgery. *Cleft Palate Journal, 26*(3), 201–208.

Guralnick, M. J. (2011). Why early intervention works: A systems perspective. *Infants Young Child, 24*, 6–28.

Guralnick, M. J. (2017). Early intervention for children with intellectual disabilities: An update. *Journal of Applied Research in Intellectual Disabilities, 30*(2), 211-229. DOI: 10.1111/jar.12233.

Riklin, E., Andover, M. S., Annunziato, R. A. (2019). The effects of society on the psychosocial functioning of those with a facial difference. *Health Psychology Report, 7*(2), 87–102. DOI: https://doi.org/10.5114/ hpr.2019.85657.

Zebrowitz, L. A., Montepare, J. M. (2008). Social Psychological Face Perception: Why Appearance Matters. *Social and Personality Psychology Compass, 2*(3), 1497-1517. DOI:10.1111/j.1751-9004.2008.00109.x.

Griffin, A., M., Langlois, J., H. (2006). Stereotype directionality and attractiveness stereotyping: Is beauty good or is ugly bad? *Social Cognition, 24*(2), 187–206.

Zebrowitz, L. A., Fellous, J. M., Mignault, A., Andreoletti, C. (2003). Trait impressions as overgeneralized responses to adaptively significant facial qualities: evidence from connectionist modeling. *Personality and Social Psychology Review, 7*, 194–215.

Zebrowitz, L. A., Rhodes, G. (2004). Sensitivity to "Bad Genes" and the Anomalous Face Overgeneralization Effect: Cue Validity, Cue Utilization, and Accuracy in Judging Intelligence and Health. *Journal of Nonverbal Behavior, 28*, 167–185.

Kramer, S., Zebrowitz, L. A., San Giovanni, J. P., Sherak, B. (1995). Infants' preferences for attractiveness and babyfaceness. IN: B.G. Bardy, R.J. Bootsma, Y. Guillard (ed.), *Studies in perception and action III* (pp. 389-392). New Jersey: Lawrence Erlbaum Associates.

Łabiszewska-Jaruzelska, F. (1995). *Ortopedia szczękowa. Zasady i praktyka.* Warsaw: PZWL.

Pluta-Wojciechowska, D. (2008). Dziecko z wadą rozwojową twarzoczaszki. IN: B. Cytowska, B. Winczura, A. Stawarski (ed.), *Dzieci chore, niepełnosprawne i z utrudnieniami w rozwoju* (pp. 331-348). Cracow: Wydawnictwo Impuls.

Mański, A. (2016a). *Dziecko z dysmorfią twarzy. Badania empiryczne uwarunkowań jego percepcji u matek i nauczycieli.* Warsaw: Difin.

Mański, A. (2016b). The image of a child with facial dysmorphism in a mother's perception. *Acta Neuropsychologica, 14*(2), 165–180.

Kowalik S. (1989). *Upośledzenie umysłowe. Teoria i praktyka rehabilitacji.* Warsaw: PWN.

Janowski, K., Steuden, S., Kuryłowicz, J., Nieśpiałowska-Steuden, M. (2009). The Disease-Related Appraisals Scale: a tool to measure subjective perception of the disease situation. IN: K. Janowski, S. Steuden (ed.), *Biopsychosocial aspects of health and disease* (pp.108-125). Lublin: CPPP Scientific Press.

Kędziora, S. (2007). Wpływ przewlekłej choroby dziecka na funkcjonowanie rodziny. *Nauczyciel i Szkoła, 3-4* (36-37), 57-66.

Liberska, H., Matuszewska, M. (2011). Model rodziny z dzieckiem niepełnosprawnym. IN: H. Liberska (ed.), *Rodzina z dzieckiem niepełnosprawnym – możliwości i ograniczenia* (pp. 42-66). Warsaw: Difin.

Pisula, E. (2012*). Rodzice dzieci z autyzmem.* Warsaw: Wydawnictwo Naukowe PWN.

Brzezińska, A. I., Kaczan, R., Ohme, M., Resler-Maj, A., Wiliński, M. (2009). *Droga do samodzielności. Jak wspomagać rozwój dzieci i młodzieży z ograniczeniami sprawności.* Gdańsk: GWP.

Kościelska M. (2000). *Oblicza upośledzenia.*Warsaw: PWN.

Pąchalska M. (2019) Integrated self system: a microgenetic approach. *Acta Neuropsychologica* 17(4):349-392.

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