

Morphological Peculiarities of Head and Face Structure in Patients with Neuro-Muscular Diseases

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The cephalometric status of patients with Congenital Cataracts Facial Dysmorphism Neuropathy syndrome (CCFDN), Hereditary Motor and Sensory Neuropathy – Lom (HMSNL) and Congenital Myasthenic Syndrome (CMS) type Ia and of healthy Gypsies is studied. The investigation includes 38 patients with CCFDN, 54 patients with HMSNL, 19 patients with CMS type Ia and 77 healthy Gypsies. The program of the study includes 20 cephalometric features and 5 cephalometric indices. Generalized the patients from both sexes showed a tendency towards less head and face measurements compared to the healthy persons. Compared to the healthy persons, the patients from both sexes have longer face, relatively less wide forehead, but larger head length than bizygomatic diameter. The male patients have relatively wider head breadth and upper face part, while the female patients have relatively bigger head length and narrower upper face part.

Key words: neuromuscular diseases, cephalometric features.

Introduction

The hereditary neuromuscular diseases are a heterogeneous group of diseases attacking the skeletal musculature and the peripheral nerves. Among these diseases are also the congenital cataracts facial dysmorphism neuropathy syndrome (CCFDN), the hereditary motor and sensory neuropathy – Lom (HMSNL) and the congenital myasthenic syndrome (CMS) type Ia. The investigated diseases are genetically determined (with autosome-recessive type of inheritance) and spread only among Gypsy families. Up to the present days a wide investigations thoroughly in medical aspect of the diseases are made [2, 3]. The aim of the present study is to analyse the inter group differences in the cephalometric status of patients with CCFDN, HMSNL and CMS type Ia and healthy Gypsies.

Material and Methods

Subject of the study are 38 patients (18 males and 20 females) with CCFDN, 54 patients (26 males and 28 females) with HMSNL, 19 patients (5 males and 14

females) with CMS type Ia and 77 healthy Gypsies (36 males and 41 females). The program of the study includes 20 cephalometric features and 5 indices (Table 1), taken by the methods of Martin, Saller [1]. The statistical significant differences are assessed by the one-way analysis of variance (ANOVA) with post hoc tests by Bonferroni and Tamhane for multiple comparisons (after the rejection of the null hypothesis by ANOVA).

Table 1. Investigated cephalometric features and indices

Cephalometrical features		Cephalometrical indices
Head length (gl-op)	Nasal-subnasal length (n-sn)	Head index
Head breadth (eu-eu)	Nasal-pronasal length (n-prn)	$I = (eu-eu) / (g-op) * 100$
Minimal frontal diameter (fl-ft)	Nose protrusion (sn-prn)	Morphological face index
Bizygomatic diameter (zy-zy)	Philtrum length (sn-sto)	$I = (n-gn) / zy-zy) * 100$
Bigonial diameter (go-go)	Labrale sup.-labrale inf. distance (ls-li)	Jugofrontal index
Trichion-nasion distance (tr-n)	Interocular diameter (en-en)	$I = (fl-ft) / (zy-zy) * 100$
Physiognomical face height (tr-gn)	Biocular diameter (ex-ex)	Transversal cephalo-facial index
Morphological face height (n-gn)	Nose breadth (al-al)	$I = (zy-zy) / (eu-eu) * 100$
Morphological upper face height (n-pr)	Lip length (ch-ch)	Jugomandibular index
Physiognomical upper face height (n-sto)	Head circumference (g-op)	$I = (go-go) / (zy-zy) * 100$

Results and Discussion

The comparative between group analyses shows a common for both sexes tendency towards smaller head and face measurements in the patients, compared to the healthy individuals (Fig. 1). This tendency is most strongly expressed in the patients with CCFDN, and most slightly – in the patients with HMSNL. Applying the dispersion analysis and suitable post hoc procedures about multiple comparison, 14 feature for men and 13 for women are distinguished in which the patients mean values differs significantly from those of the healthy persons (Table 2). The differences are more strongly expressed to a large degree in women than in men. For both sexes more frequent are the significant differences between healthy individuals and CCFDN patients than between healthy individuals and HMSNL and CMS type Ia patients.

Significant differences of the cephalometric indexes' characteristics in the healthy and diseased individuals are available. The data about head index (Fig. 2)

Table 2. Statistical significant differences between investigated subgroups of patients and healthy persons

Groups		Features																		
		head circum.	g-op	eu-eu	fl-ft	tr-n	zy-zy	go-go	tr-gn	n-gn	n-sio	sto-gn	tr-sn	al-al	n-prn	sn-prn	sn-sto	ls-li	ch-ch	en-en
Males	healthy / CCFDN	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*
	healthy / HMSNL						*	*	*		*								*	
	healthy / CMS						*						*						*	
	CCFDN / HMSNL					*													*	
	CCFDN / CMS																			
	HMSNL / CMS												*						*	
Females	healthy / CCFDN	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*
	healthy / HMSNL	*					*					*			*				*	
	healthy / CMS						*					*			*				*	
	CCFDN / HMSNL	*	*			*	*	*	*	*	*	*	*	*	*	*	*	*	*	*
	CCFDN / CMS		*		*	*	*	*	*	*	*	*	*	*	*	*	*	*	*	*
	HMSNL / CMS						*					*			*			*	*	*

*Statistical significant differences (P<0.05)

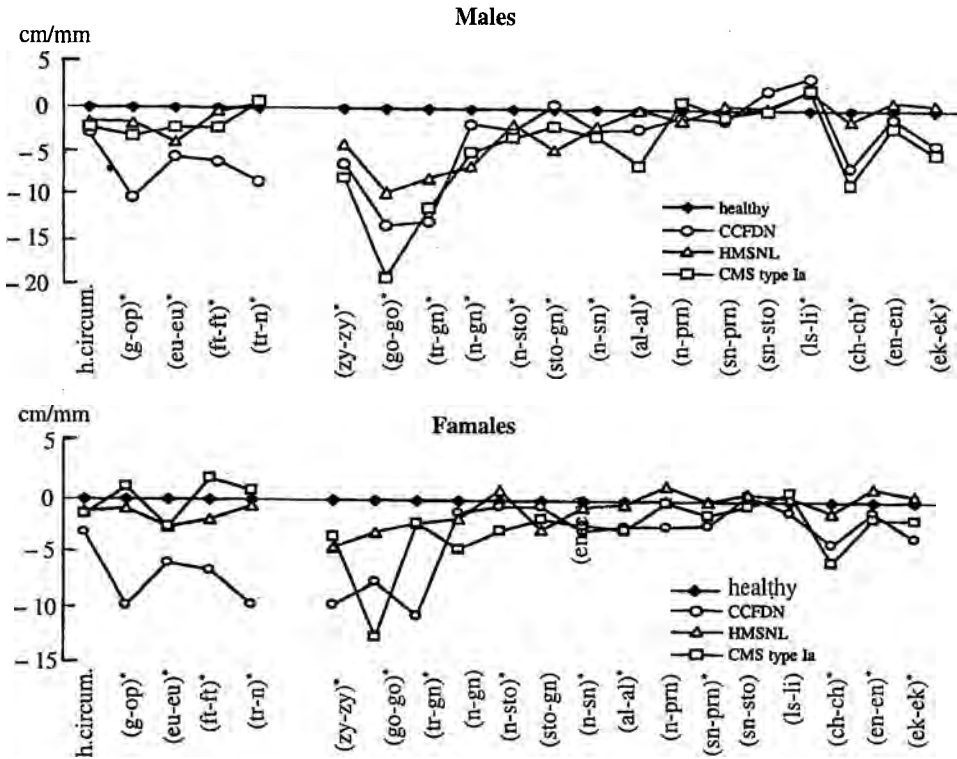


Fig. 1. Differences between mean values of the investigated cephalometric features in patients with CCFDN, HMSNL, CMA type Ia and healthy persons

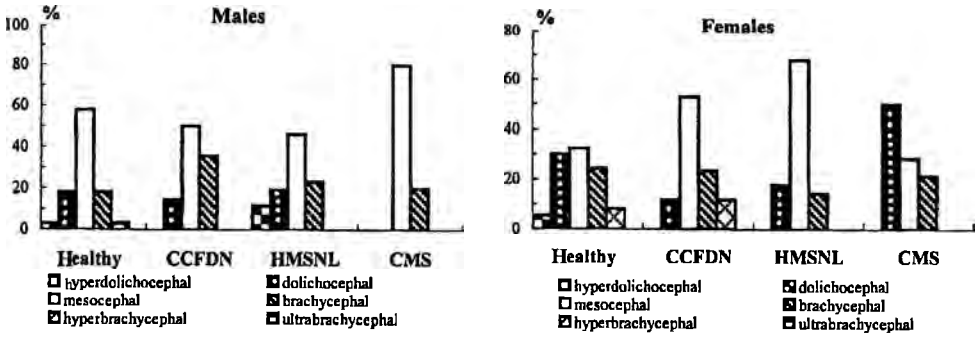


Fig. 2. Distribution of the investigated patients and healthy persons by head index categories

show that males and females with CCFDN, as well as the males with CMS type Ia have relatively higher head width while the males and females with HMSNL and the females with CMS type Ia have relatively higher head length. The CCFDN males basically distinguish by higher frequency of brachycephal forms, the HMSNL males — by higher frequency of dolichocephal and hyperdolichocephal ones, and the CMS

type Ia males — by very high frequency of mesocephal, as well as, by total absence of dolichocephal and hyperdolichocephal forms. As the CCFDN females basically distinguish by higher frequency of mesocephal and lower frequency of dolichocephal forms, the HMSNL females — by lower frequency of brachycephal forms and higher frequency of mesocephal forms while the CMS type Ia females — by higher frequency of dolichocephal forms. Analyzing the jugofrontal index in the patients from both sexes, a tendency for relatively higher forehead width compared to the bizygomatic diameter is established (Fig. 3). In the male and female patients, the frequency of very wide forms is higher, and the frequency of the wide forms are near or less to those in the healthy ones. The data about transversal cephalo-facial index show that the patients from both sexes have relatively less bizygomatic diameter compared to the head length (Fig. 4). A tendency of higher frequency about middle and below middle forms than those about over middle and large forms is observed. According to the data about jugomandibular index, the CCFDN, HMSNL and CMA type Ia males, as well as the CMS type Ia females have brighter upper face part in relation to the lower part (Fig. 5). Opposite, the CCFDN and HMSNL females have relatively narrower upper face part than the lower one. In the ill males and the CMS type Ia females is established a lower frequency of short and very short forms, and

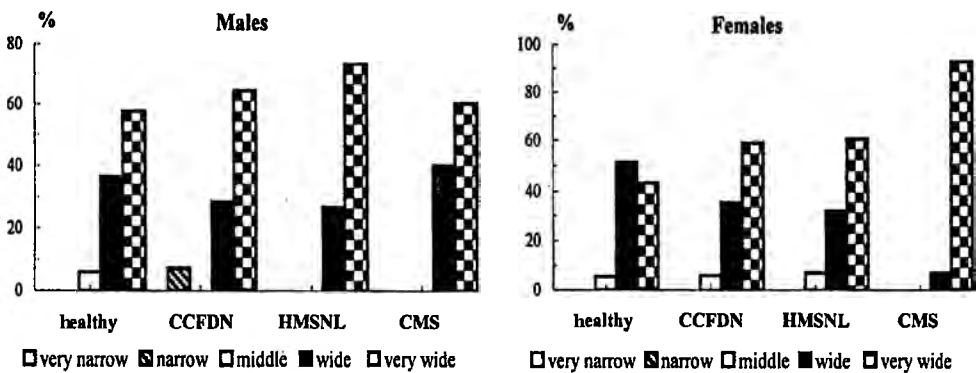


Fig. 3. Distribution of the investigated patients and healthy persons by jugofrontal index categories

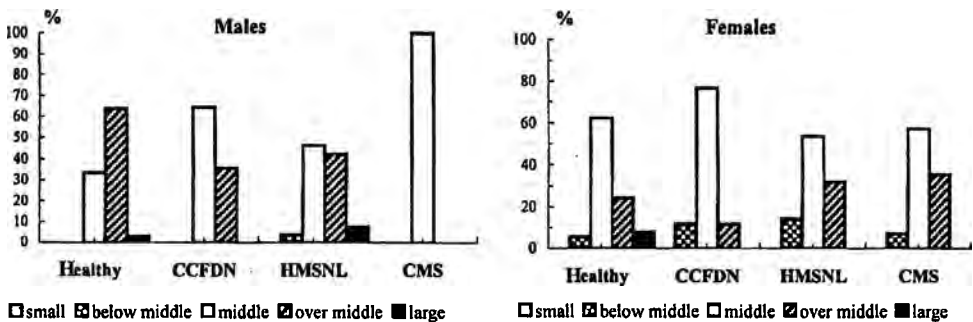


Fig. 4. Distribution of investigated patients and healthy persons by transversal cephalo-facial index categories

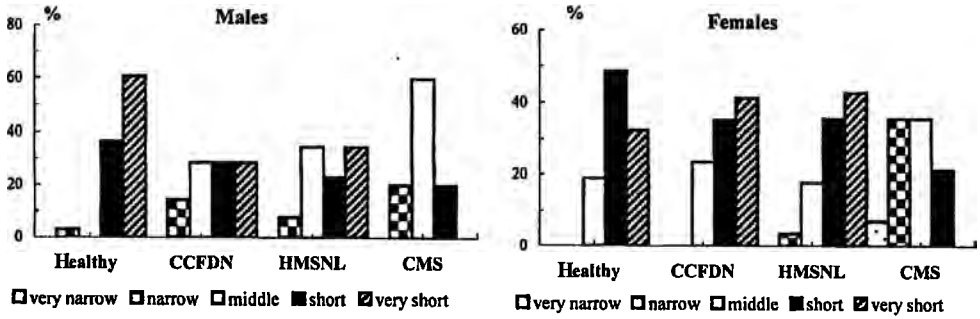


Fig. 5. Distribution of investigated patients and healthy persons by jugomandibular index categories

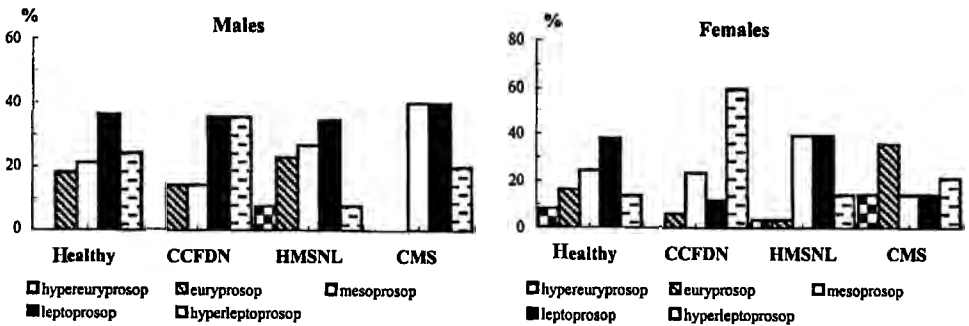


Fig. 6. Distribution of investigated patients and healthy persons by morphological face index categories

considerably higher frequency of middle and narrow forms. The HMSNL and CCFDN females have a higher frequency of very short forms. For the patients from both sexes, a tendency toward longer faces as a whole, and separately for the upper face part is established. According to morphological face index data, the patients more frequently are mesoprosopes, leptoprosopes and hyperleptoprosopes, and less frequently euryprosopes and hypereuryprosopes (Fig. 6).

Conclusions

A common tendency towards less head and face measurements in the patients from both sexes compared to the healthy persons is available. This tendency is strongly expressed in the CCFDN patients and slightly — in the HMSNL ones. In males reliable differences are established only between the HNSNL and CCFDN patients, as well as between the HMSNL and CSM type Ia patients. For females reliable differences are established between all subgroups of patients.

Compared to the healthy individuals, the patients from both sexes have longer face, relatively less forehead width, but larger head length than bizygomatic diameter. The male patients have relatively wider head breadth, while the female patients have relatively bigger head length.

References

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