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## NUTRITIONAL STANDARD FOR CHILDREN WITH OROFACIAL CLEFTS

### POSTĘPOWANIE ŻYWIENIOWE U DZIECI Z ROZSZCZEPEM CZĘŚCI TWARZOWEJ CZASZKI

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#### **Abstract**

**Introduction:** Treatment of children with orofacial clefts is a multi-stage process, usually extending over many years and requiring intervention of numerous specialists. Most of the problems in such children before the tissue reconstruction surgery are related to feeding and airway protection during swallowing. Feeding of children with orofacial clefts is the more difficult the more severe the defect is. Such children are at an increased risk of body weight deficit and malnutrition.

**Aim of the study:** The aim of the study is to present the nutritional standard for children with orofacial clefts.

**Feeding principles for children with orofacial clefts:** If the clinical state of the child and the emotional state of the mother allow, children with orofacial clefts should be breastfed or bottle-fed with breast milk. If feeding with breast milk is not possible, children should receive appropriate formulas for infants. Their diet can usually be expanded at the same time as in healthy infants and should comply with the nutritional model or standard for children aged 6-12 months. Various feeding techniques are used in children with orofacial clefts, depending not only on the type of the defect, but also the experience of the institution taking care of the child. Such children may require a diet with higher calories due to their increased energy expenditure related to eating. In the case of body weight deficit and/or malnutrition resulting from inadequate diet, a change of the feeding technique should be considered, and, subsequently, a diet modification. The modification may mean an extra formula feeding (in children fed with breast milk) or earlier introduction of supplementary foods. Sometimes a different feeding method than oral feeding must be used, e.g. through a naso-gastric tube or, in extreme cases, a feeding stoma.

It is of utmost importance that infants with the said defects gain the optimal body weight before the planned operation, since malnutrition may be a significant reason for delaying the planned operation; it may also have an adverse impact on the healing of wounds and increase the risk of postoperative complications.

**Conclusions:** Infants with birth defects in the form of orofacial clefts are at risk of developmental disorders and, in particular, body weight deficit.

Systematic assessment of their nutritional status aims at identifying potential irregularities, defining their underlying reasons and implementing an appropriate treatment.

In the therapy of children with orofacial clefts, an individualised, comprehensive approach to nutrition, in line with recommendations of the team taking care of the child, is of utmost importance.

**Key words:** cleft lip and palate, feeding

#### **Streszczenie**

**Wstęp:** Leczenie dziecka z wadą rozszczepową twarzoczaszki jest wieloetapowe i jest to zazwyczaj proces wieloletni, wymagający zaangażowania wielu specjalistów. U dzieci tych do czasu zabiegu rekonstrukcji tkanek najczęściej problemów związanych jest z karmieniem oraz przeciwdziałaniem przedostawaniu się pokarmu do dróg oddechowych. Karmienie dzieci z rozszczepem jest tym trudniejsze im większy jest zakres wady. Dzieci te znajdują się w grupie zwiększonego ryzyka wystąpienia niedoboru masy ciała i niedożywienia.

**Cel pracy:** Celem pracy jest przedstawienie schematu postępowania żywieniowego u dzieci z rozszczepem części twarzowej czaszki.

**Zasady dotyczące żywienia dziecka z rozszczepem części twarzowej czaszki:** Jeśli pozwala na to stan kliniczny dziecka i stan emocjonalny matki zaleca się karmienie dzieci z rozszczepami twarzoczaszki poprzez przystawienie do piersi lub żywienie pokarmem odciągniętym. Gdy karmienie naturalne jest niemożliwe dzieci te powinny otrzymywać odpowiednie mieszanki przeznaczone do karmienia niemowląt. Rozszerzanie ich diety zazwyczaj jest możliwe w tym samym czasie co u zdrowych niemowląt i powinno odbywać się zgodnie z modelem lub schematem żywienia dzieci w drugim półroczu życia. U dzieci z rozszczepem twarzoczaszki stosowane są różne techniki karmienia, co warunkowane jest nie tylko rodzajem wady, ale także doświadczeniami ośrodka opiekującego się dzieckiem. Dzieci te mogą wymagać zwiększonej kaloryczności diety z uwagi na większy wydatek energetyczny związany z przyjmowaniem pokarmu. W przypadku niedoboru masy ciała i/lub zdiagnozowanego niedożywienia wynikającego z nieadekwatnej do potrzeb diety, u dzieci tych rozważać należy zmianę techniki karmienia, następnie sposób modyfikacji diety. Modyfikacja dotyczyć może podjęcia decyzji o dokarmianiu sztucznym (u dzieci żywionych naturalnie) czy też wcześniejszym wprowadzeniu żywności uzupełniającej. Czasem konieczne jest zastosowanie alternatywnej dla doustnej drogi karmienia np. poprzez zgłębnik czy też wyjątkowo przetokę odżywczą.

Uzyskanie, przez niemowlęta z omawianymi wadami, optymalnej masy ciała do czasu planowanego zabiegu operacyjnego jest bardzo ważne, ponieważ niedożywienie może być jednym z istotnych czynników odpowiedzialnych za odrzucanie terminu planowanego zabiegu, może niekorzystnie wpływać na gojenie się ran oraz zwiększać ryzyko wystąpienia powikłań pooperacyjnych.

**Wnioski:** Niemowlęta z wrodzonymi wadami rozszczepowymi części twarzowej czaszki znajdują się w grupie ryzyka wystąpienia zaburzeń rozwoju, a szczególnie niedoborów masy ciała.

Systematyczna ocena stanu odżywienia ma na celu określenie potencjalnych nieprawidłowości, zdefiniowanie ich przyczyn oraz wdrożenie odpowiedniego postępowania.

W postępowaniu terapeutycznym u dzieci z wadami rozszczepowymi twarzoczaszki ważne jest indywidualne, kompleksowe podejście do żywienia, zgodnie z zaleceniami zespołu opiekującego się dzieckiem.

**Słowa kluczowe:** rozszczep wargi i podniebienia, żywienie

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## INTRODUCTION

Treatment of children with orofacial clefts is a multi-stage process, usually extending over many years. The complex consequences of clefts require the involvement of numerous specialists, such as surgeon, neonatologist/paediatrician, orthodontist, phoniatriest, speech therapist, psychologist and other specialists, if necessary, in the therapeutic process. Depending on their severity, clefts result in deformation of lips, nose and facial skeleton and in impairment of such functions as sucking, swallowing, breathing, speech and hearing (1).

One of the essential elements of the treatment is surgical correction of the defect to reconstruct the tissues in the place of the cleft. Immediately after birth, every child with an orofacial cleft should receive individualised recommendations for optimal therapeutic procedure. Prenatal identification of the defect facilitates the therapy. Most of the problems encountered in such children in the first period of their life are related to feeding and airway protection during eating.

## AIM

The aim of the study is to present the nutritional standard for children with orofacial clefts.

Description of feeding principles for children with orofacial clefts

In the therapy of children with orofacial clefts, an individualised, comprehensive approach to feeding, in line with the recommendations of the team taking care of the child, is of utmost importance (fig. 1). The feeding standard before the original reconstructive surgery may be divided into three stages:

- before surgical intervention,
- during surgical intervention,
- directly after surgical intervention.

Breast milk should be the main food for children in their first year of life. Feeding with breast milk is beneficial both for the mother and for the baby. It has been proven that breastfed children with cleft lip and palate less frequently suffer from otitis media with effusion than the formula-fed infants (2). In addition, according to Erkkilä A. T. et al, longer breastfeeding of children with such defects follows with better school performance in the future (3).

Therefore, if the clinical state of the child and the emotional state of the mother allow, such infants should be breastfed or bottle-fed with breast milk, using an individualised feeding technique. The nutritional value of breast milk allows to satisfy energy needs of the child in the first 6 months of its life and the demand for all

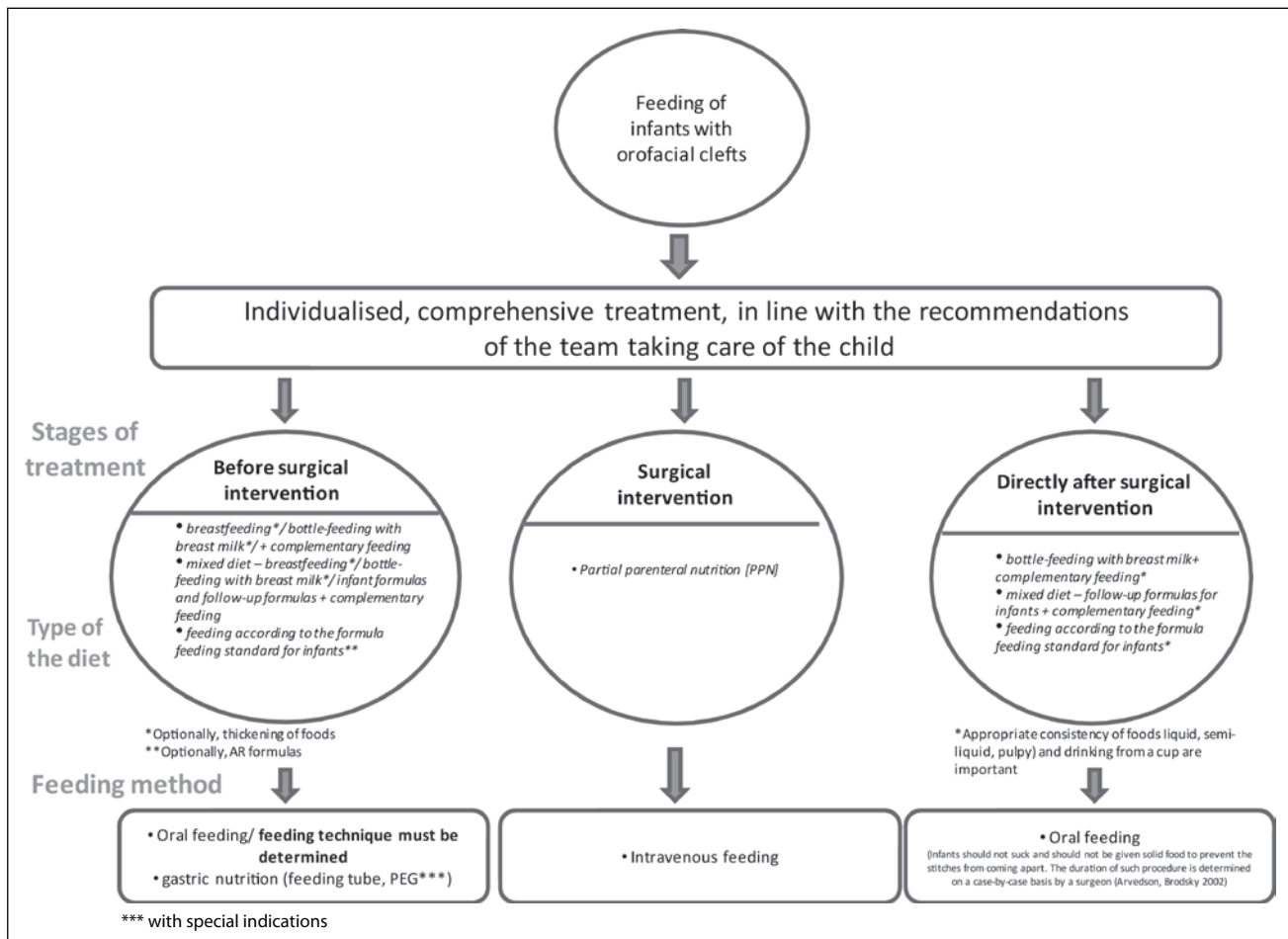


Fig. 1. Nutritional standard for children with orofacial clefts.

Ryc. 1. Schemat postępowania żywieniowego u dziecka z rozszczepem części twarzowej czaszki.

nutrients, excluding vitamin D and vitamin K. These vitamins must be provided additionally as supplements, in line with the guidelines (4-6).

Breastfeeding of children with clefts is not always possible. Ineffective sucking from the breast due to the child's defect and inappropriate milk extraction from the breast for bottle-feeding may lead to cessation of lactation. In addition, the stress of the mother due to ineffective feeding, and also the stress related to frequent visits to numerous specialists with the child, as well as hospitalisation of the child, are other factors predisposing to cessation of lactation.

When feeding with breast milk is impossible, children should be fed with appropriate infant formula or follow-up formula, in line with the formula feeding standard for infants (7, 8). Children with orofacial defects usually eat smaller amounts / smaller portions of food which is beneficial, since some of them suffer from pathological gastro-esophageal reflux (9). Therefore, they need to be fed more often/eat more meals but with smaller food portions than healthy infants at the same age. Some children with clefts may benefit from adding infant and follow-up antireflux formulas (AR-type).

After 6 months of age, the child's diet must be appropriately expanded to include products increasing its energy and nutritional value (vegetables, fruit, lean meat, fish, egg yolk, fats - olive oil, low or non-erucic acid rapeseed oil, fresh butter, cereal products/groats, iron-enhanced gruels). Children with orofacial defects are usually ready for their diet to be expanded at the same time as healthy infants (10).

Tables I and II present nutritional standards and recommendations for healthy children from various age groups which should serve as the basis for planning the diet.

Infants with cleft defects may have higher energy needs than their healthy peers. This is due to higher energy expenditure related to eating (11). The components of a higher energy diet, rich in protein from milk and milk products, meat, poultry, fish and eggs, should be determined on a case-by-case basis by the physician taking care of the child or by a dietician supervised by a physician.

In the case of body weight deficit and/or malnutrition resulting from inadequate diet, a change of the feeding technique should be considered, and, subsequently, a

Table I. Recommended nutrients profile in children's diets (4).

Tabela I. Zalecany profil składników pokarmowych w dietach dzieci (4).

Part A. Recommendations of nutrients and energy intake for infants, children and adolescents.

Część A. Zalecenia dziennego spożycia podstawowych składników pokarmowych i energii dla niemowląt, dzieci i młodzieży.

Age [years] Wiek [lata]	Body weight [kg] Masa ciała [kg]	Energy [kcal/d] Energia [kcal/d]	Protein Białko				Carbohydrates Węglowodany					
			Estimated Average Requirement (EAR) Średnie zapotrzebo- wanie (EAR)		Recommended Dietary Allowances (RDA) Zalecane spożycie (RDA)		Adequate Intake (AI) wystarczające spożycie (AI)		Recommended Dietary Allowances (RDA) Zalecane spożycie (RDA)		Adequate Intake (AI) Wystarczające spożycie (AI)	
			g/kgm.c/d	g/os/d	g/kgm.c/d	g/os/d	g/kgm.c/d	g/os/d	g/os/d	g/os/d		
Infants Niemowlęta												
0-0,5	6.5	600					1.52*	10		60		
0,5-1	9	700					1.60*	14		95		
Children Dzieci												
1-3	12	1000	0.97	12	1.17	14			130			
4-6	19	1400	0.84	16	1.10	21			130			
7-9	27	1800	0.84	23	1.10	30			130			
Boys Chłopcy												
10-12	38	2400	0.84	32	1.10	42			130			
13-15	53	3000	0.84	45	1.10	58			130			
16-18	67	3400	0.81	54	0.95	64			130			
Girls Dziewczęta												
10-12	37	2100	0.84	31	1.10	41			130			
13-15	51	2450	0.84	43	1.10	56			130			
16-18	56	2500	0.79	44	0.95	53			130			

diet modification. The modification may mean an extra formula feeding (in children fed with breast milk) or earlier introduction of supplementary foods. Sometimes a different feeding method than oral feeding must be used, e.g. through a naso-gastric tube or, in extreme cases, a feeding stoma (11).

#### DETAILED NOTES ON THE NUTRITIONAL STANDARD

Feeding of children with orofacial clefts is the more difficult the more severe the defect is. Due to impaired functioning of such structures as tongue and/or larynx and/or pharynx, children with such defects are at risk of problems with swallowing, pathological gastro-esophageal reflux, as well as apnea and food aspiration. Dysphagia in those infants is usually caused by numerous factors (12).

Children with cleft lip, similarly to some children with minor cleft soft palate or submucous cleft palate, may be effectively breastfed. However, this requires finding the optimal position for the child during feeding and providing consultation to the mother in the lactation clinic, if any problems occur (13).

Since the connection between oral cavity and nasal cavity in more pronounced palate clefts is open, children with such defects are unable to suck breast, since they cannot generate a negative pressure in the oral cavity. Such children should be bottle-fed with breast milk.

Various feeding techniques are used in children with orofacial clefts, depending not only on the type of defect, but also on the experience of the institution taking care of the child. Appropriate bottles and individually chosen teats are used for feeding of infants with clefts. The best position for bottle-fed children is a half-vertical

Part B. Recommendations of vitamins intake for infants, children and adolescents.

Część B. Zalecane dzienne spożycie witamin dla niemowląt, dzieci i młodzieży.

Vitamins Witaminy	Units Jednostki		Age [years] Wiek [lata]										
			Infants Niemowlęta		Children Dzieci			Boys Chłopcy			Girls Dziewczęta		
			0-0,5	0,5-1	1-3	4-6	7-9	10-12	13-15	16-18	10-12	13-15	16-18
A	µg retinol equivalnet µg równoważnika retinolu	AI	400	500									
		EAR			280	300	350	450	630	630	430	490	490
		RDA			400	450	500	600	900	900	600	700	700
D <sub>3</sub>	µg cholecalciferol µg cholekalcyferolu	AI	5	5	5	5	5	5	5	5	5	5	5
D <sub>3</sub> *	µg cholecalciferol µg cholekalcyferolu	AI	10	10									
		EAR			10**	10	10	10	10	10	10	10	10
		RDA			15**	15	15	15	15	15	15	15	15
E	mg tocopherol equivalnet mg równoważnika tokoferolu	AI	4	5	6	6	7	10	10	10	8	8	8
K	µg phyloquinone µg filochinonu	AI	5	10	15	20	25	40	50	65	40	50	55
B <sub>1</sub>	mg thiamin mg tiaminy	AI	0.2	0.3									
		EAR			0.4	0.5	0.7	0.9	1.0	1.0	0.8	0.9	0.9
		RDA			0.5	0.6	0.9	1.0	1.2	1.2	1.0	1.1	1.1
B <sub>2</sub>	mg riboflavin mg ryboflawiny	AI	0.3	0.4									
		EAR			0.4	0.5	0.8	0.9	1.1	1.1	0.8	0.9	0.9
		RDA			0.5	0.6	0.9	1.0	1.3	1.3	1.0	1.1	1.1
PP	mg niacin equivalnet mg równoważnika niacyny	AI	2	4									
		EAR			5	6	9	9	12	12	9	11	11
		RDA			6	8	12	12	16	16	12	14	14

\*Institute of Medicine, Dietary Reference Intake for Calcium and Vitamin D, Food and Nutrition Board, Nat. Acad. Press, Washington, 2010

\*\*Dobrzańska A. i wsp. Normy żywienia zdrowych dzieci w 1-3. roku życia – stanowisko Polskiej Grupy Ekspertów. Część II – Omówienie poszczególnych składników odżywczych. Standardy Medyczne 2012, 9: 200-205.

or vertical position which prevents choking and food getting into the nasal cavity and Eustachian tube. It is also important that the bottle be held at an angle and then vertically during feeding to prevent the child from swallowing excessive amount of air and that the infant be held in a vertical position for a sufficiently long time after feeding (14, 15).

In children with a significant loss of palate, individually adjusted dental apparatus, the so-called palatal plates may be used for effective bottle-feeding. The efficiency of feeding using such apparatus differs according to different authors (16-20). Turner et al found that the use of palatal plates in both breastfed and bottle-fed children with cleft lip and palate reduces the feeding times and

Part C. Recommendations of minerals intake for infants, children and adolescents.

Część C. Zalecane dzienne spożycie składników mineralnych dla niemowląt, dzieci i młodzieży.

Minerlas Składniki mineralne	Units Jednostki		Age [years] Wiek [lata]										
			Infants Niemowlęta		Children Dzieci			Boys Chłopcy			Girls Dziewczęta		
			0-0,5	0,5-1	1-3	4-6	7-9	10-12	13-15	16-18	10-12	13-15	16-18
Calcium [Ca] Wapń [Ca]	mg	AI	200	260									
		EAR			500	800	800	1100	1100	1100	1100	1100	1100
		RDA			700	1000	1000	1300	1300	1300	1300	1300	1300
Phosphorus [P] Fosfor [P]	mg	AI	150	300									
		EAR			380	410	500	1050	1050	1050	1050	1050	1050
		RDA			460	500	600	1250	1250	1250	1250	1250	1250
Magnesium [Mg] Magnez [Mg]	mg	AI	30	70									
		EAR			65	110	110	200	340	340	200	300	300
		RDA			80	130	130	240	410	410	240	360	360
Iron [Fe] Żelazo [Fe]	mg	AI	0,3										
		EAR		7	3	4	4	7	8	8	7(8)*	8	8
		RDA		11	7	10	10	10	12	12	10(15)*	15	15
Zinc [Zn] Cynk [Zn]	mg	AI	2										
		EAR		2.5	2.5	4	4	7	8.5	8.5	7	7.3	7.3
		RDA		3	3	5	5	8	11	11	8	9	9
Copper [Cu] Miedź [Cu]	mg	AI	0.2	0.3									
		EAR			0.25	0.3	0.5	0.5	0.7	0.7	0.5	0.7	0.7
		RDA			0.3	0.4	0.7	0.7	0.9	0.9	0.7	0.9	0.9

\*Źródło: Jarosz M. [red.]: Normy żywienia dla populacji polskiej – nowelizacja. Instytut Żywności i Żywienia, Warszawa 2012

Table II. Contribution of energy from protein, fat and carbohydrates in balanced children's diet in relation to their age.

Tabela II. Udział energii z białka, tłuszczu i węglowodanów w zrównoważonej diecie dziecka w zależności od wieku.

Age Wiek	Protein [%] Białko [%]	Fat [%] Tłuszcz %	Carbohydrates [%] Węglowodany %
0-12 months breastfeeding formula fed infants 0-12 m.ż. karmienie naturalne karmienie sztuczne	9 12.5	41 35	50 53
2-3 years 2-3 r.ż.	12.0	32	56
4-6 years 4-6 r.ż.	12.0	32	56
7-9 years 7-9 r.ż.	12.0	no less than 31 nie mniej niż 31	57
10-12 years and more 10-12 r.ż. i powyżej	12.0	no less than 31 nie mniej niż 31	57

Źródło: Jarosz M. [red.]: Normy żywienia dla populacji polskiej – nowelizacja. Instytut Żywności i Żywienia, Warszawa 2012

increases the volume consumed at a shorter time (19). According to Ize et al, children with cleft lip and palate, fed both with a syringe and with a cup / spoon, consume more food at a shorter time than the children who are breastfed or bottle-fed using a palatal plate (20).

Due to controversies regarding the impact of palatal plates not only on the effectiveness of feeding, but also on development, growth and formation of cleft jaw, we do not use this method in our centre. This is also caused by our own observations (21).

Kogo et al did not find any significant differences in weight gain of children with cleft lip and palate who were bottle-fed and fed with a cup/spoon and therefore recommended bottle-feeding as a simpler technique (22). Shaw et al believe that bottle-feeding allows to achieve satisfactory weight gains and is less time-consuming (23).

In infants with cleft palate, and in particular in those with combined defects, who experience significant problems when fed orally, a naso-gastric tube may be used (24). However, such procedure used for a long time may be one of the reasons for feeding problems in subsequent months or years of the child's life (25). It is of utmost importance that infants with the said defects obtain the optimal body weight before the planned surgery, since malnutrition may be one of significant factors for delaying the date of the planned operation; it may also have an adverse impact on the healing of wounds and increase the risk of postoperative complications. This requires longer hospitalisation and thus generates higher costs.

Feeding problems demonstrated by body weight deficit in the pre-operative period concern 32% of infants with unilateral cleft lip, 30-40% with unilateral cleft lip and palate and approximately 50% with isolated cleft palate. In the case of some combined defects, the percentage is as high as 90-100% (26). The unpublished study carried out at the Children and Youth Surgery Department of the Institute of Mother and Child showed that among patients with orofacial clefts operated in 2012, the percentage of children with body weight deficit (<15c) amounted to 23.8% .

In children and youth with orofacial clefts, the incidence of dental problems is higher than in the general population, they also have occlusal abnormalities (27).

Some of them require orthodontic treatment i.e. permanent dental braces for their dental and occlusal disorders.

Energy and nutritional value of the diets of children and youth wearing permanent dental braces should not differ from general recommendations for the population of this age (tab. I, II). Consistency of food is important, since consumption of hard foods may damage the braces. Hard foods, such as fruit and vegetables, should be eaten ground, also after heat treatment. Consumption of stone fruit should also be reduced, since their parts may be hard to remove from the braces. The diet should include a reduced amount of sweet juices and products high in carbohydrates, in particular monosaccharides and disaccharides, which is important for prevention of dental caries.

## CONCLUSIONS

Infants with birth defects in the form of orofacial clefts are at risk of developmental disorders and, in particular, body weight deficit.

Systematic assessment of the nutritional status of such children is particularly important to identify potential irregularities and their reasons and to start an appropriate treatment.

In the therapy of children with orofacial clefts, an individualised, comprehensive approach to nutrition, in line with recommendations of the team taking care of the child, is of utmost importance.

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