



Orofacial function of persons having Wolf-Hirschhorn syndrome

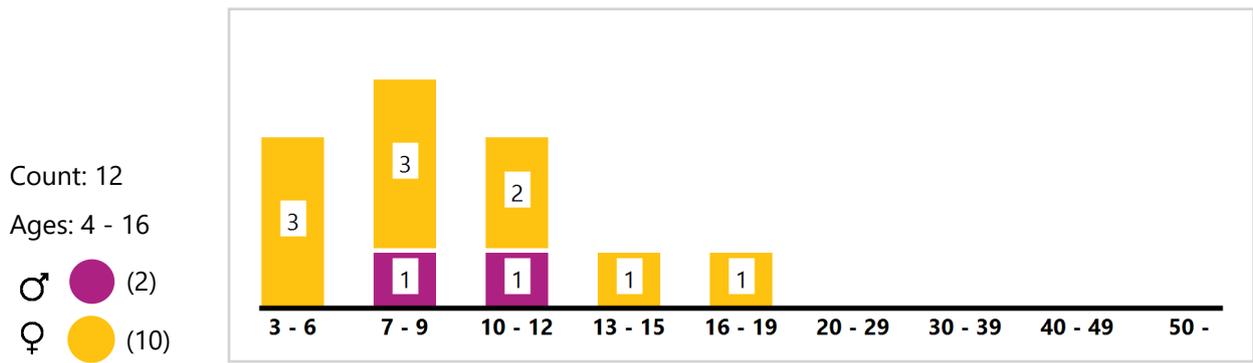
Report from observation charts



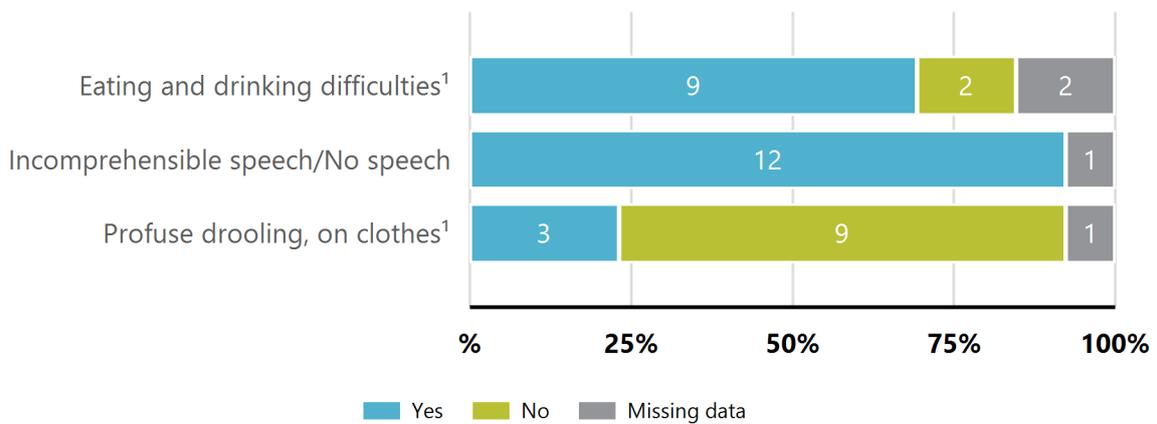
12 observation charts

Synonym	Monosomy 4p syndrome, 4p deletion syndrome, Wolf's syndrome
ICD-10	Q93.3
Estimated occurrence	2:100,000 live births.
General symptoms	Newborn babies with monosomy 4p weigh slightly less than what is normal. Muscle weakness (hypotonia) is common. The palate is sometimes cleft. Everyone with this diagnosis suffers from a developmental disability which is most often severe. Eye abnormalities may be present. Abnormalities of the heart, often uncomplicated, are present in 50% of the children; 40% have hearing defects. It is common for these children to be short. Most suffer from epilepsy which is often difficult to treat. Some children are unable to straighten out their fingers, which are sometimes bent to the side, and large birth marks can be present. There is an increased risk of infection, especially stomach and bowel infections.
Oral symptoms	Characteristic facial features are associated with the diagnosis. Muscle weakness and a possible cleft palate often make it difficult for infants to suckle. It is common for the palate to be high, and the jaw and teeth to be small. Speech and communication difficulties are common, as are eating difficulties and drooling.
Oral treatment	<ul style="list-style-type: none"> • If there is a cleft palate, a specialist team is responsible for follow-up and treatment. • It is important for people with Wolf-Hirschhorn syndrome to have contact with dental care early on for strengthened preventive care and oral health information. • Oral motor training and stimulation is often urgently needed in cases with eating difficulties. • There is often cause for speech, language and communication training. • Contact with a physician is recommended before dental treatment for medical conditions, such as heart defects.
Sources	The MHC database Rare diseases Dokumentation-Ågrenska

Age distribution



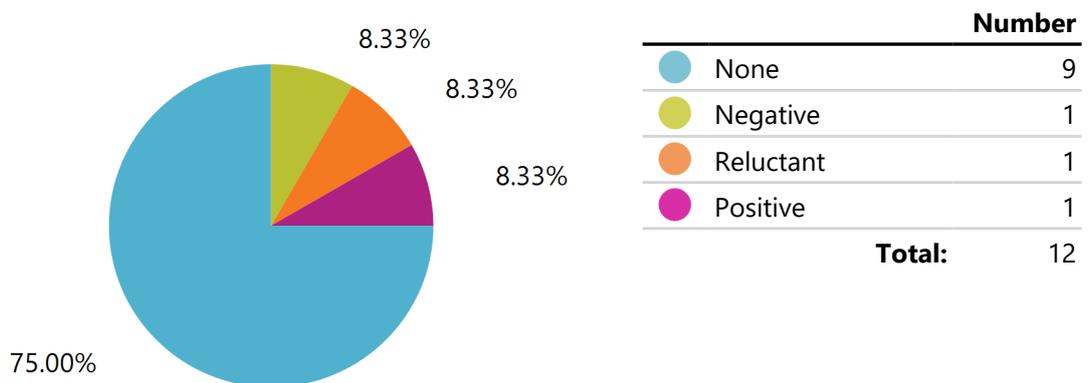
Summary



Certain caution should be observed when interpreting the chart because the number of individuals is less than 100.

¹ Reported via questionnaire

Acceptance of dental examination



Oral health

Caries

		3-6 years	7-12 years	13-19 years
deft	Examined	3	7	
Number of carious or filled deciduous teeth	Number of individuals with deft=0	3	7	
	Mean			
	Standard deviation			
	No data	3	7	
DMFT	Examined		7	2
Number of carious or filled permanent teeth	Number of individuals with DMFT=0		7	2
	Mean			
	Standard deviation			
	No data		7	2

Oral health index (indices)¹

	0	1	2	3	4	5	6	Missing data	Number
Calculus	1	0	0	1	0	0	1	9	12
Gingivitis	1	0	0	0	0	0	0	11	12
Plaque	1	0	0	0	0	0	0	11	12
Tooth wear	1	0	0	0	0	0	0	11	12

CALCULUS

Calculus index is based on the presence of visible calculus on the buccal surface of 6 index teeth. 0 indicates that there is no calculus at all, 6 indicates calculus on all index teeth.

GINGIVITIS

Gingivitis index is based on the presence of visible gingivitis on the buccal surface of 6 index teeth. 0 indicates that there is no bleeding, 6 indicates bleeding on all index teeth.

PLAQUE

Plaque index is based on the presence of visible plaque on the buccal surface of 6 index teeth. 0 indicates that there is no plaque, 6 indicates plaque on all index teeth.

TOOTH WEAR

Tooth wear index is a weighted summary of the degree of tooth wear on 6 different segments. Tooth wear is only evaluated in the permanent dentition, not in the primary teeth. The final index score is based on the degree of tooth wear found in most segments.

0: No tooth wear or minor wear of enamel in either of the segments.

1: Marked tooth wear of the enamel, possibly exceeding into dentin.

2: tooth wear in the dentine reaching up to 1/3 of the tooth crown.

3: Tooth wear in the dentine reaching up to more than 1/3 of the tooth crown. If 3 is given in any segment then SI is 3.

¹ This variable was introduced in version 2 (2008) of the Observation chart.

Occlusal relationship

	Number
Neutral bite	1
Overbite	2
Underbite	0
Missing data	9
Total:	12

Maximum jaw opening

Children younger than 10 years

	Number
-20	0
21-30	0
31-40	0
41-50	0
51-	0
Missing data	7
Total:	7

Children, 10 years or older, and adults

	Number
-20	0
21-30	0
31-40	0
41-50	0
51-	0
Missing data	5
Total:	5

Profile¹

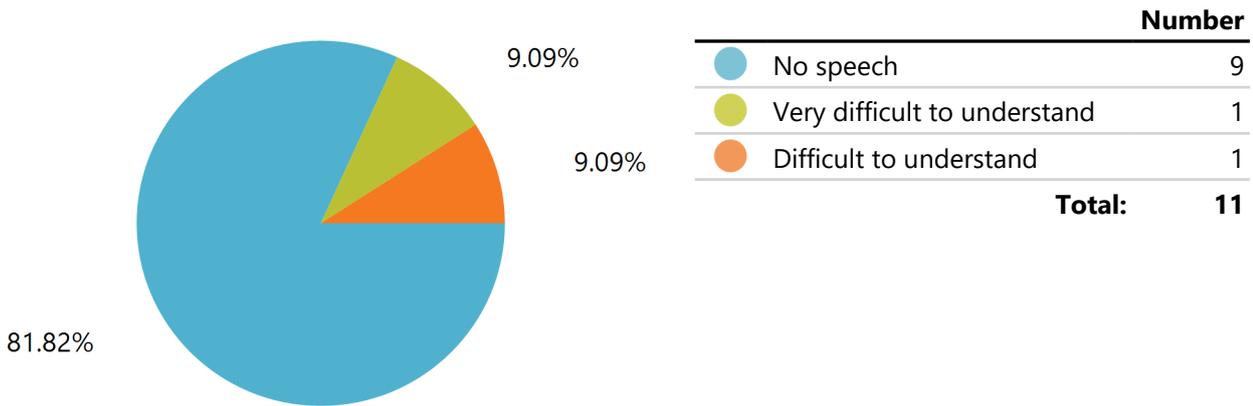
	Number
Normal	3
Convex	7
Concave	1
Missing data	1
Total:	12

Mandibular plane¹

	Number
Normal	5
Increased	0
Reduced	4
Missing data	3
Total:	12

¹ This variable was introduced in version 2 (2008) of the Observation chart.

Speech problems



Clinical findings

Number of yes-answers

	Total = 12 (%)	♂ = 2 (%)	♀ = 10 (%)	Missing Data
Low muscle tone in lips	5 (42)	1 (50)	4 (40)	1
High palate	5 (42)	1 (50)	4 (40)	5
Impaired tongue motility	5 (42)	0 (0)	5 (50)	6
Spacing	4 (33)	1 (50)	3 (30)	7
Open mouth at rest	2 (17)	1 (50)	1 (10)	1
Low muscle tone in tongue	2 (17)	1 (50)	1 (10)	4
Narrow palate	2 (17)	0 (0)	2 (20)	6
Over crowding	2 (17)	1 (50)	1 (10)	8
Reduced stability in neck	2 (17)	0 (0)	2 (20)	1
Low muscle tone in masticatory muscles	1 (8)	1 (50)	0 (0)	8
Deep bite with gingival contact	1 (8)	1 (50)	0 (0)	10
Short tongue frenulum	1 (8)	1 (50)	0 (0)	5