Orofacial function of persons having
Optic nerve hypoplasia

Report from questionnaires | 13 questionnaires

Synonym Septo-optic dysplasia, ONH syndrome

ICD-10 Q04.4

Estimated occurance 7:100,000 children aged 0-19

General symptoms Optic nerve hypoplasia can occur as a single diagnosis or in combination with other types of hormonal and/or neurological disruptions or behavioural disorders. Effect on vision varies from mild to severe on one eye or both. Symptoms normally occur during the first year of life. There is nothing in the information we have today to say that vision will get worse with time, but rather that the damage is static. Hormonal deficiencies are common in children with this diagnosis, and most need hormone supplements. It is important to get an early diagnosis to avoid the hormonal deficiencies causing too much damage. Variation between the different functional impairments is great. Development of fine and gross motor skills is often affected. Other symptoms that may occur are epilepsy, paralysis of one side of the body, developmental disability, behavioural disorder and autism.

Oral symptoms In cases with a neurological functional impairment, oral motor skills and speech development can be affected.

Oral treatment

- Early contact with dental services for intensified prophylactic care and oral hygiene information is essential in cases of difficulty with dental treatment and tooth brushing.
- Regular check-ups of dental and jaw development. Orthodontist should be consulted when needed.
- Training in oral motor skills and extra stimulation in cases of eating disorders, speech difficulties and impaired saliva control may be necessary.
- Speech and language difficulties should be treated by a speech therapist.

Sources The MHC database Rare diseases Dokumentation-Ågrenska

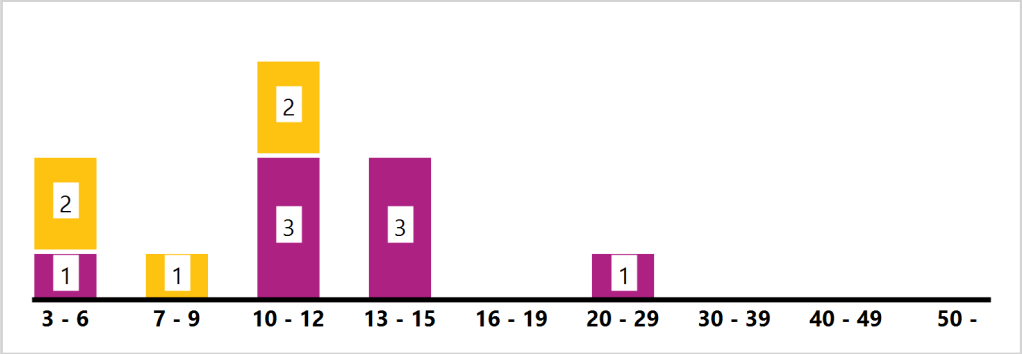
Age distribution

Number: 13

Ages: 5 - 27

♂ (8)

♀ (5)



Additional diagnoses

Medical impairment	Yes	No	Missing data
Inborn heart defect	1	12	0
Other cardiovascular disease	0	13	0
Epilepsy	2	10	1
Asthma	2	11	0
Need of respiratory support	0	13	0
Allergy	3	10	0

Neuropsychiatric diagnosis	Yes	No	Missing data
ADHD/ADD	1	11	1
Autism (Includes autism, Asperger syndrome and autistic traits)	9	2	2

General disability	Yes	No	Missing data
Intellectual disability	2	6	5
Motoric functional impairment	4	3	6
Visual impairment	1	0	12
Hearing loss	0	13	0
Communication difficulties	2	5	6

About dental care and oral health

Do you feel that you receive the dental care you need?	Number
Yes, very much so	10
Yes, somewhat	2
Missing data	1
Total:	**13**

How many times per year do you normally seek dental care?	Number
Less than once per year	3
One time per year	4
Two times per year	3
Three or more times per year	3
Total:	**13**

When were your teeth last X-rayed?	Number
During the past two years	8
More than two years ago	2
Never had my teeth X-rayed	3
Total:	**13**

Do you look after your teeth in a good way?	Number
Yes, very much so	7
Yes, somewhat	5
No, not really	1
Total:	**13**

Who brushes your teeth?	Number
I always brush myself	3
Someone else always helps me	8
Sometimes I brush myself	2
Total:	**13**

How often are your teeth brushed?	Number
Once per day	2
Two times per day	10
Three or more times per day	1
Total:	**13**

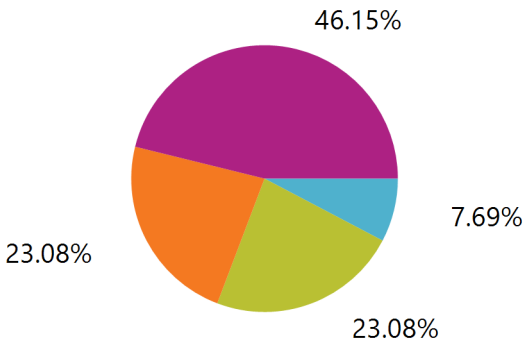
About dental care and oral health

	Yes	No	Missing data
Does your mouth hurt?	0	12	1
Does your mouth feel dry?	2	10	1
Have you ever taken a serious hit to your permanent front teeth?	1	12	0
Do you feel that you have a divergent bite?	5	7	1
Have you had a brace?	1	12	0
Do you feel that you need orthodontics/a brace?	1	8	4

Do you grind or press your teeth at night?	Number
Never	9
Once or twice per week	1
Every night	3
Total:	**13**

Do you grind or press your teeth during the day?	Number
Never	9
Once or twice per week	2
Every night	2
Total:	**13**

About eating



Do you have any problems with eating?	Number
Yes, very much so	1
Yes, somewhat	3
No, not really	3
No, not at all	6
Missing data	0
Total:	**13**

	Yes	No	Missing data
Do you cough daily in connection with meals?	1	12	0
Do you gag daily in connection with meals?	2	11	0
Do you get acid reflux daily?	2	10	1
Do you throw up often (at least twice per week)?	0	13	0
Do you have a poor appetite?	0	13	0
Does it take a long time before you can swallow a mouthful?	0	10	3
Do you press your tongue forward when you swallow so that food ends up outside the mouth?	0	10	3
Do you find it difficult to chew, i.e. grind food using your molars?	2	10	1
Do you find it difficult to take food from the spoon using your lips?	2	10	1
Have you had problems with food and drink leaking out through the corners of your mouth?	1	9	3
Does food tend to remain in your mouth after meals?	1	11	1
Do you get nutrition in any other way than through your mouth?	0	13	0

About drooling

Do you drool?	Number
Never drool	9
Drool sometimes – not every day	2
Drool often – every day	2
Missing data	0
Total:	**13**

How much do you drool?	Number
Slight drooling, only on the lips	1
Moderate drooling, on lip and chin	2
Very profuse drooling	1
Total:	**4**

Is your drooling a problem for you?	Number
Yes, somewhat	1
No, not really	1
No, not at all	2
Total:	**4**

Is your drooling a problem for your family or people around you?	Number
Yes, somewhat	2
No, not at all	2
Total:	**4**