



## Orofacial function of persons having Arthrogryposis Multiplex Congenita Report from observation charts

The survey comprises 40 observation charts.

**Estimated occurrence:** 1:4 500 live births.

**Etiology:** The etiology may be classified as:

- Muscular causes (various congenital muscular diseases).
- Neurogenic causes (deformities of the central nervous system, loss of motor neurons in the spinal marrow, loss of peripheral nerves).
- Connective tissue diseases
- Reduced mobility of the fetus owing to insufficient space in the uterus (lack of amniotic fluid, twin pregnancy, heart-shaped uterus).
- Maternal illness during pregnancy.

**General symptoms:** Stiffness in several joints is the common denominator. The disease is not progressive; on the contrary treatment may result in improvement. It is common for these children to be born with one dislocated hip and a club foot.

**Orofacial/odontological symptoms:** The jaw-joints may also be affected, which results in a restricted ability to open the jaws. An overjet and open bite in the region of the front teeth, a narrow, high palate and a small lower jaw all occur. Many individuals have restricted oral motor skills, which affects their ability to suck, chew, swallow, and speak. Drooling and teeth grinding occur frequently.

**Orofacial/odontological treatment:**

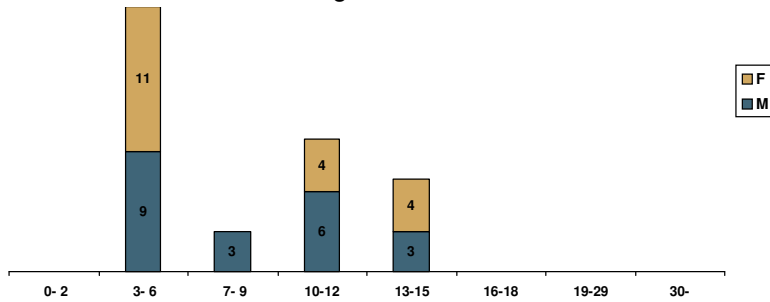
- When the jaws do not open properly, the function of the jaw joint should be investigated, and appropriate treatment thereafter prescribed (such as jaw-stretching exercises or surgery).
- An orthodontist should be consulted between the ages of 7 and 9 in order to identify dental aberrations or malocclusions and to plan any necessary orthodontic treatment to correct malocclusions.
- Training in oral motor skills and extra stimulation in cases of eating disorders, speech difficulties and drooling may be necessary.
- Teeth grinding should be followed up, and be managed with a splint when necessary.

**Sources:**

The MHC database - The Mun-H-Center database of orofacial manifestations in rare diseases.

The Newsletter of the Ågrenska Center.

Age distribution

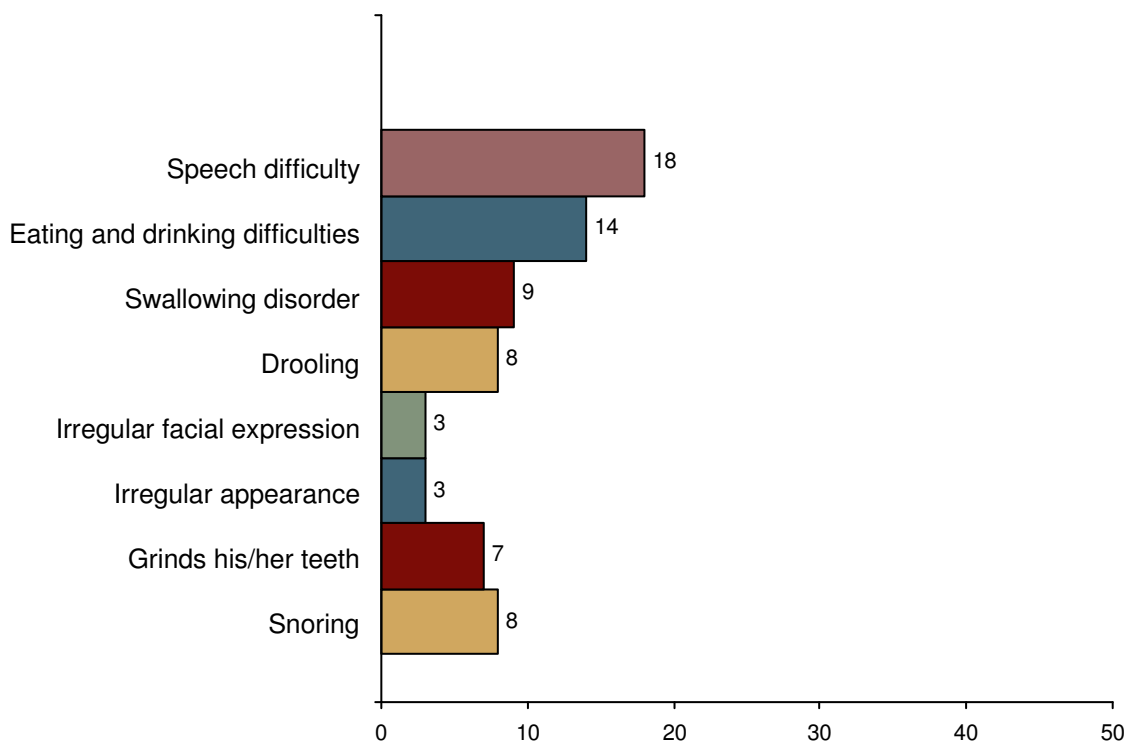


Number: 40

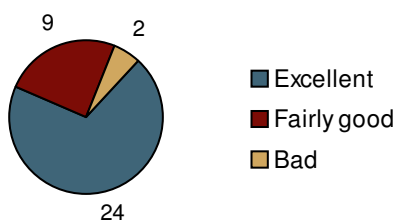
Ages: 3 -- 15 years

Sex: M (21) + F (19)

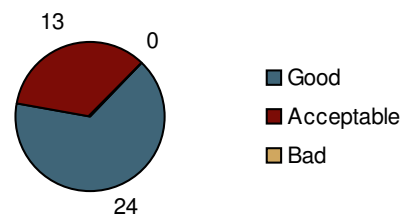
Orofacial problems



Oral health

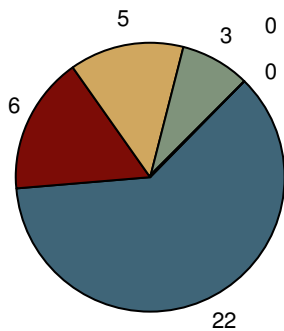


Oral hygiene



## Behaviour in the treatment situation

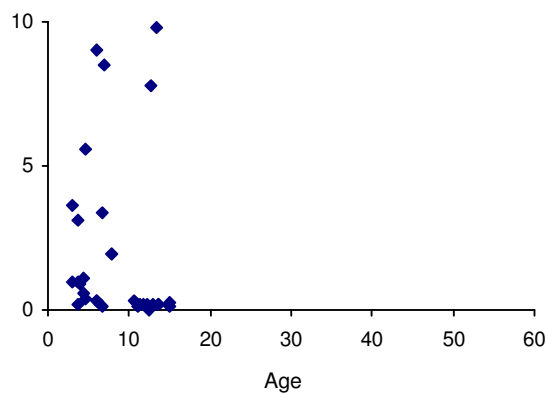
How calm and co-operative is the patient at time of examination?



- (22) ■ Examination is possible without problem
- (6) ■ Examination is possible without problems, some reaction is observed
- (5) ■ Examination can continue if adjusted to patient's reactions
- (3) ■ Reactions are considerable and examination is obviously affected
- (0) ■ Examination is practically impossible to complete
- (0) ■ Patient refuses examination

How does the patient cope with treatment in general?

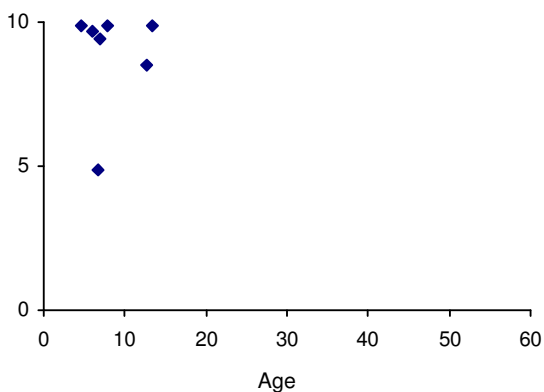
0=no problems/10=great problems



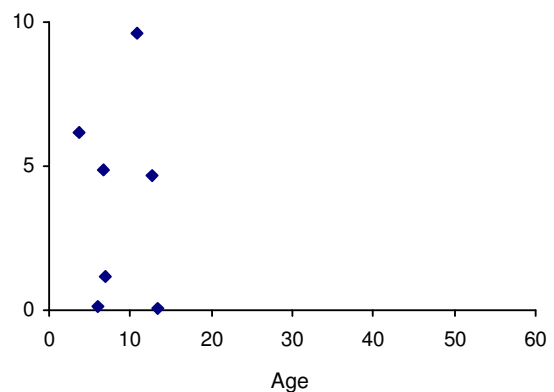
If there are treatment problems:

To what degree are the problems, if any, due to NN's handicap? To what degree are the problems, if any, due to NN's fear?

0=not at all/10=to a very high degree



0=not at all/10=to a very high degree



| <b>Clinical findings</b>          | <b>Total<br/>N=40</b> | <b>Boys/Men<br/>N=21</b> | <b>Girls/Women<br/>N=19</b> | <b>Not<br/>evaluated</b> |
|-----------------------------------|-----------------------|--------------------------|-----------------------------|--------------------------|
| Speech difficulty                 | 19                    | 12                       | 7                           | 0                        |
| Open mouth at rest                | 14                    | 10                       | 4                           | 0                        |
| M mentalis is overactive          | 11                    | 6                        | 5                           | 1                        |
| Gingivitis                        | 10                    | 7                        | 3                           | 1                        |
| Narrow palate                     | 10                    | 7                        | 3                           | 2                        |
| Drooling                          | 9                     | 7                        | 2                           | 0                        |
| High palate                       | 9                     | 6                        | 3                           | 2                        |
| Hypomineralisation                | 9                     | 4                        | 5                           | 2                        |
| Post normal bite                  | 9                     | 4                        | 5                           | 5                        |
| Reduced mobility in TMJ           | 9                     | 5                        | 4                           | 1                        |
| Reduced mobility in tongue        | 9                     | 8                        | 1                           | 1                        |
| Horizontal over-bite 6 mm or more | 8                     | 4                        | 4                           | 4                        |
| Low muscle tone in lower lip      | 8                     | 6                        | 2                           | 0                        |
| Frontal open bite                 | 7                     | 6                        | 1                           | 4                        |
| Low muscle tone in upper lip      | 7                     | 5                        | 2                           | 0                        |
| Mask-like expression              | 7                     | 5                        | 2                           | 0                        |
| Over crowding                     | 7                     | 6                        | 1                           | 4                        |
| Abrasion - insignificant          | 6                     | 5                        | 1                           | 3                        |
| Deviation on opening              | 6                     | 4                        | 2                           | 2                        |
| Mouth breathing                   | 6                     | 5                        | 1                           | 3                        |
| Reduced mobility in soft palate   | 6                     | 4                        | 2                           | 9                        |
| Upper jaw seems small             | 6                     | 4                        | 2                           | 2                        |
| Upper lip is inactive and raised  | 6                     | 4                        | 2                           | 0                        |
| Cross bite                        | 5                     | 2                        | 3                           | 3                        |
| Dental trauma                     | 5                     | 1                        | 4                           | 4                        |
| Grinds his/her teeth              | 5                     | 2                        | 3                           | 1                        |
| Low muscle tone in tongue         | 5                     | 5                        | 0                           | 1                        |
| Lower jaw seems small             | 5                     | 3                        | 2                           | 2                        |
| Other oral habits                 | 5                     | 3                        | 2                           | 0                        |
| Proclined upper incisors          | 5                     | 4                        | 1                           | 3                        |
| Retroclined lower incisors        | 5                     | 4                        | 1                           | 4                        |
| Enamel hypoplasia                 | 4                     | 3                        | 1                           | 1                        |
| Hyper-sensitive in oral cavity    | 4                     | 4                        | 0                           | 2                        |
| Lower lip is flaccid and inactive | 4                     | 3                        | 1                           | 0                        |
| Pre normal bite                   | 4                     | 3                        | 1                           | 4                        |
| Reduced stability in neck         | 4                     | 3                        | 1                           | 1                        |
| Spacing                           | 4                     | 4                        | 0                           | 3                        |
| Deep bite with gingival contact   | 3                     | 1                        | 2                           | 4                        |
| Edge to edge bite                 | 3                     | 2                        | 1                           | 3                        |
| Facial asymmetry                  | 3                     | 2                        | 1                           | 0                        |
| Misshaped occlusal plane          | 3                     | 1                        | 2                           | 4                        |
| Philtrum seems long               | 3                     | 2                        | 1                           | 0                        |
| Philtrum seems short              | 3                     | 2                        | 1                           | 0                        |
| Posturing forward                 | 3                     | 2                        | 1                           | 4                        |
| Reduced mobility in neck          | 3                     | 3                        | 0                           | 1                        |