

Occlusion and orofacial functions in Beckwith-Wiedemann syndrome - a pilot study

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Background

Beckwith-Wiedemann syndrome (BWS) is a rare disorder that may affect a range of organs, especially during early childhood. Macroglossia is common (figure 1). This may cause dentoskeletal aberrations (figure 2) and affect orofacial functions. Hemihypertrophy occurs in one fourth of the cases. Tongue reduction surgery may improve development of the jaws (figures 3 and 4). The aim of this pilot study was to examine occlusion and oral function in a group of patients with the diagnosis.



Figure 1.



Figure 2.



Figure 3.



Figure 4.

Methods

We invited 34 patients registered with Beckwith-Wiedemann syndrome to participate in a pilot study. Dental occlusion was analysed using photographs and dental casts.

Nordic Orofacial Test- Screening (NOT-S) was used for evaluation of 12 domains of orofacial functions. A total score from 0 to 12 was given with 0 indicating no dysfunction.

Results

Eleven patients (age 4-30 years, median 11; 4 males) accepted to participate. The main findings regarding occlusion are presented in tables 1 and 2. Orthodontic treatment had been performed in one of the tongue-operated patients with permanent dentition.

Table 1. Occlusion in unoperated patients with BWS

| | Mixed dentition (n=3) | Permanent dentition (n=3) | Total (n=6) |
|---------------------------|-----------------------|---------------------------|-------------|
| Underbite | 1 | 1 | 2 |
| Edge-to-edge relationship | 0 | 1 | 1 |
| Vertical open bite | 3 | 3 | 6 |
| Displaced midline | 2 | 3 | 5 |
| Lateral crossbite | 2 | 2 | 4 |

Table 2. Occlusion in patients with BWS who had undergone tongue reduction surgery

| | Primary dentition (n=2) | Mixed dentition (n=1) | Permanent dentition (n=2) | Total (n=5) |
|---------------------------|-------------------------|-----------------------|---------------------------|-------------|
| Underbite | 1 | 1 | 0 | 2 |
| Edge-to-edge relationship | 0 | 0 | 0 | 0 |
| Vertical open bite | 2 | 0 | 0 | 2 |
| Displaced midline | 1* | 0 | 2 | 3 |
| Lateral crossbite | 0 | 0 | 1 | 1 |

* Missing data on one patient

NOT-S total scores were 1-6 (median 2) in unoperated participants and 3-6 (median 4) in tongue-operated individuals. Distribution of the scores in the different NOT-S domains for the eleven patients are shown in figure 5. The tongue was resting behind the teeth in six of the patients, between the teeth in three cases and between the lips in two. Speech deviations mainly occurred as interdentalization.

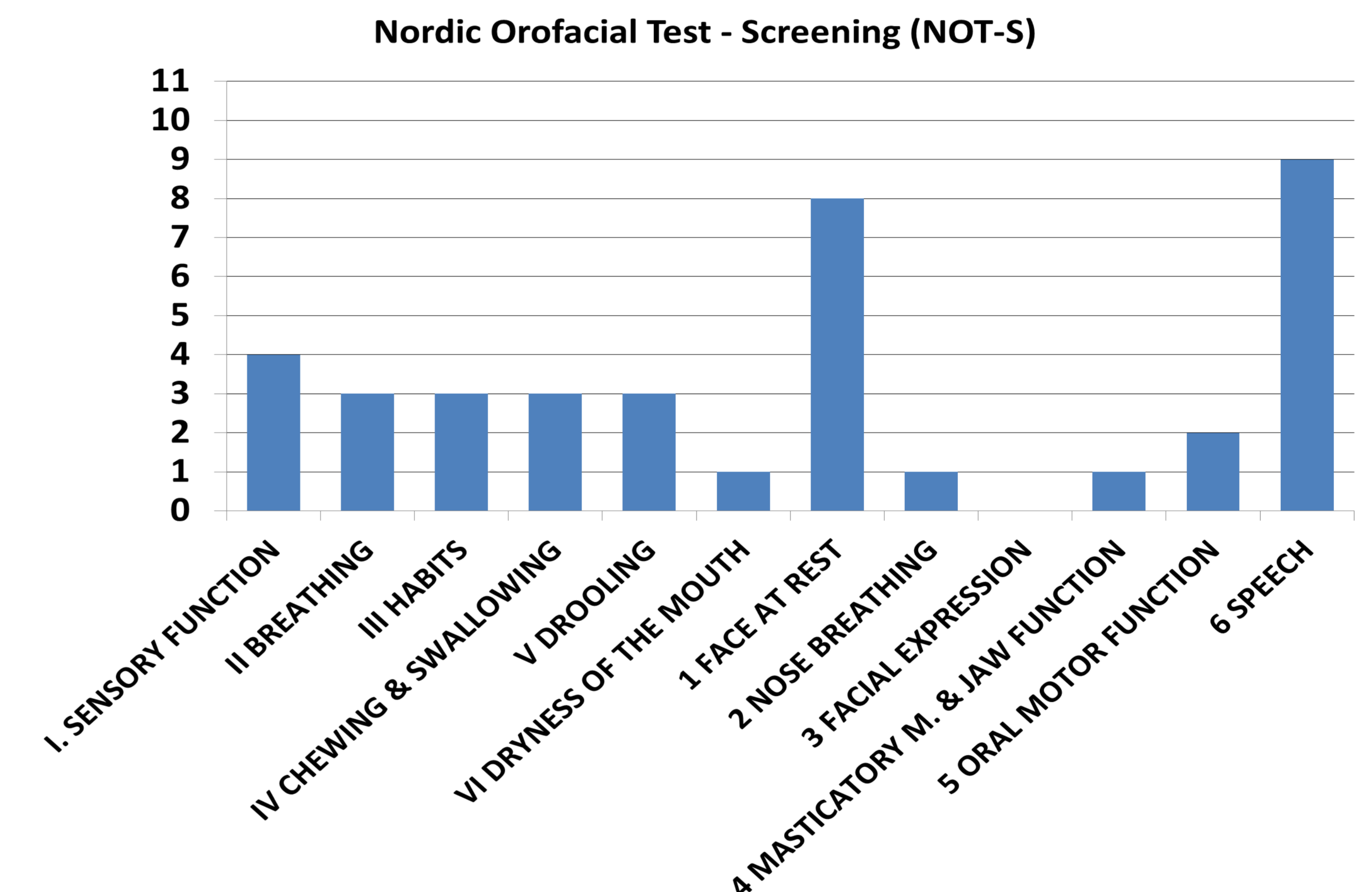


Figure 5.

Conclusions

- Vertical open bite and crossbite seem to be more frequent in unoperated individuals with BWS
- Underbite occurs in some unoperated and operated patients
- Deviating facial resting position and altered articulation were frequent irrespective of prior surgery