

Validated Oral Health Indices in Bone Marrow Failure Syndromes

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Background

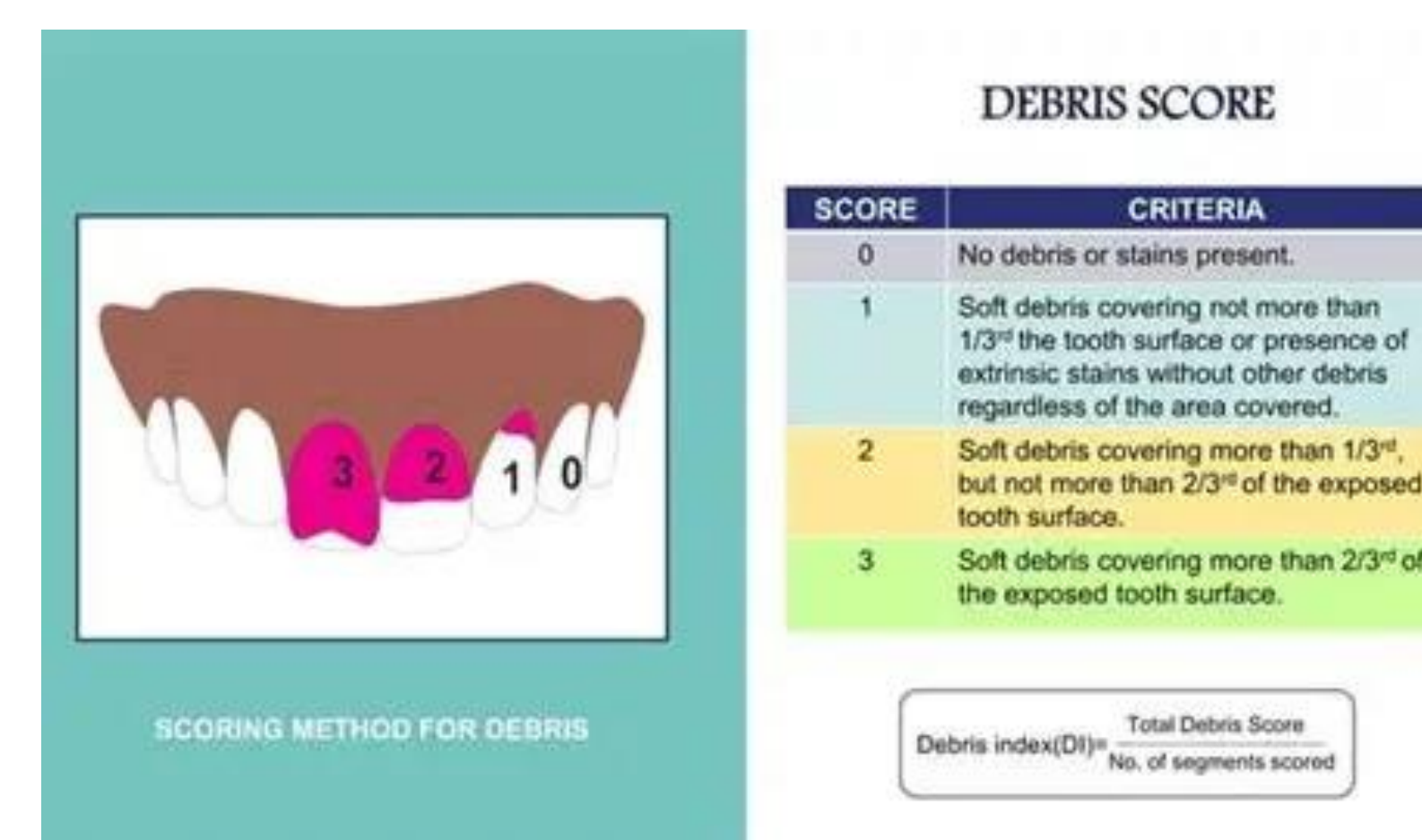
- Fanconi Anemia, Schwachman- Diamond Syndrome, Dyskeratosis Congenita, and Telomere Biology Disorders can be classified under the umbrella term 'Inherited Bone Marrow Failure Disorders' (IBMFD).
- According to the National Cancer Institute, the inherited bone marrow failure syndromes (IBMFS) are a group of rare genetic blood disorders in which there is usually some form of aplastic anemia (failure of the bone marrow to produce blood), associated with a family history of the same disorder.
- These patients have a higher risk for development of either leukemias or solid tumors. According to the Cancer Foundation, patients with Fanconi anemia are at a potentially 700-fold increased risk for head and neck squamous cell carcinoma.

Objective

The primary objective of this pilot study was to expand the knowledge of bone marrow failure disorders, namely Fanconi Anemia (FA), Schwachman-Diamond Syndrome (SDS), Dyskeratosis Congenita (DKC), and Telomere Biology Disorders (TBD) using three standardized oral indices.

Methods

This IRB-approved case cohort study involved assessment of patients with IBMFS by one of two calibrated examiners using: Simplified Oral Health Index (OHI-S), Modified Gingival Index (MGI), The Ulcer Severity Score (USS/OMAS), and a Mean Oral Erythema score. Clinically visible dental carious lesions were also recorded. Subjects were recruited from the Cancer & Blood Diseases Institute Clinic at Cincinnati Children's Hospital Medical Center.



Simplified Oral Hygiene Index.

Score	Inflammation	Appearance
0	Normal	None
1	Mild inflammation	Slight changes in color and texture, but not in all portions of gingival marginal or papillary
2	Mild inflammation	Slight changes in color and texture in all portions of gingival marginal or papillary
3	Moderate	Bright surface inflammation, erythema, edema, and/or hypertrophy of gingival marginal or papillary
4	Severe inflammation	Erythema, edema, and/or marginal gingival hypertrophy of the unit or spontaneous bleeding, papillary, congestion, or ulceration

Modified Gingival Index.

Lesion Number Score	Signs
0	No lesions
1	1-2 localised lesions
2	3-5 localised lesions
3	6-10 lesions
4	>10 lesions or diffuse (or very large) lesions

Lesion Severity Score	Signs
0	No lesion
1	Appears superficial (only mucosa missing)
2	Deeper structures involved (greater depth than No. 1)
3	Multiple lesions and variable severity (1, 2 and/or 4)
4	Same as 2 and has active appearance (active = hyperaemic and/or darkened lesion crater)
5	Same as 4 plus active haemorrhage or adherent blood clot

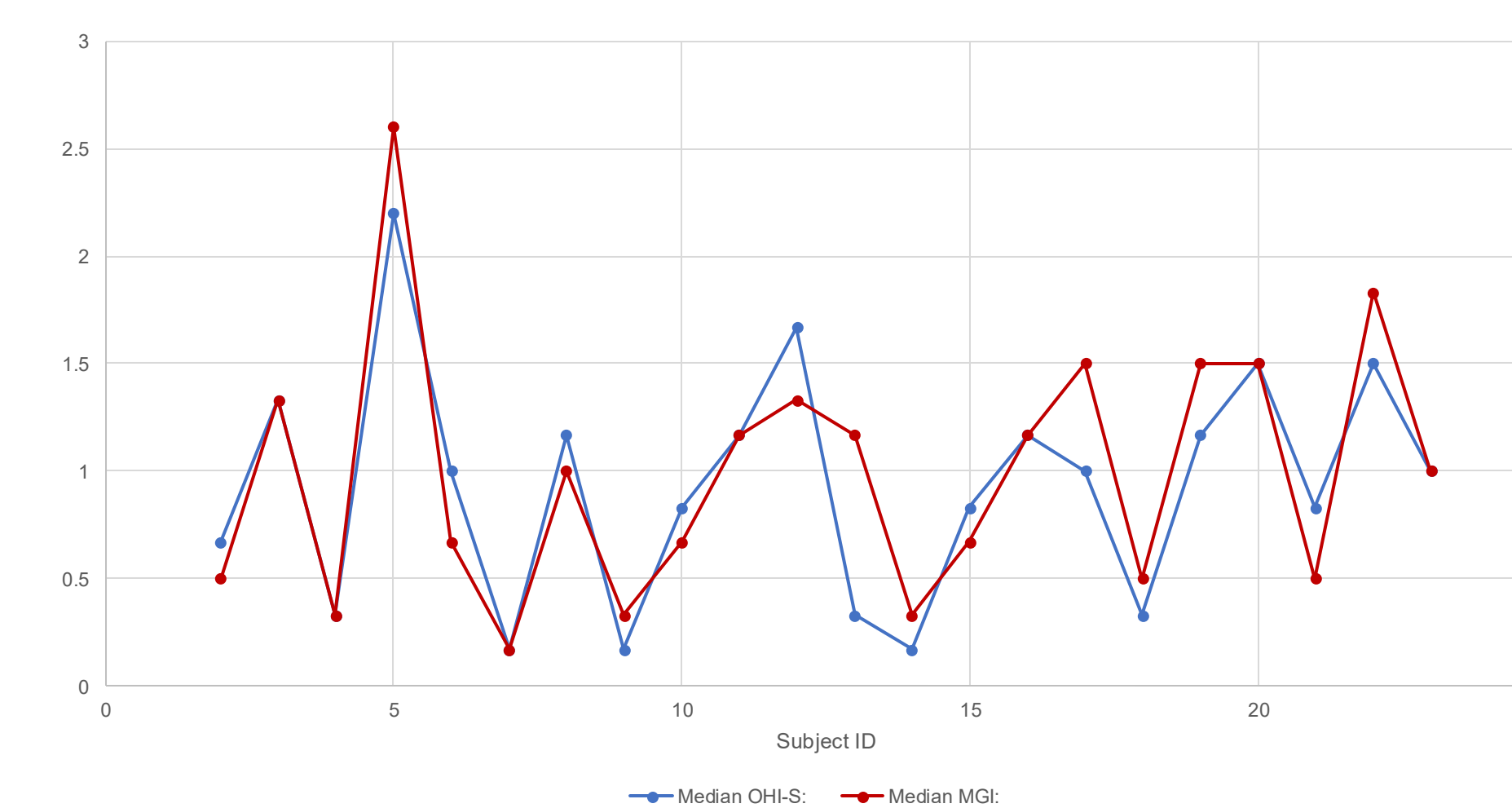
Ulcer Severity Score

Results

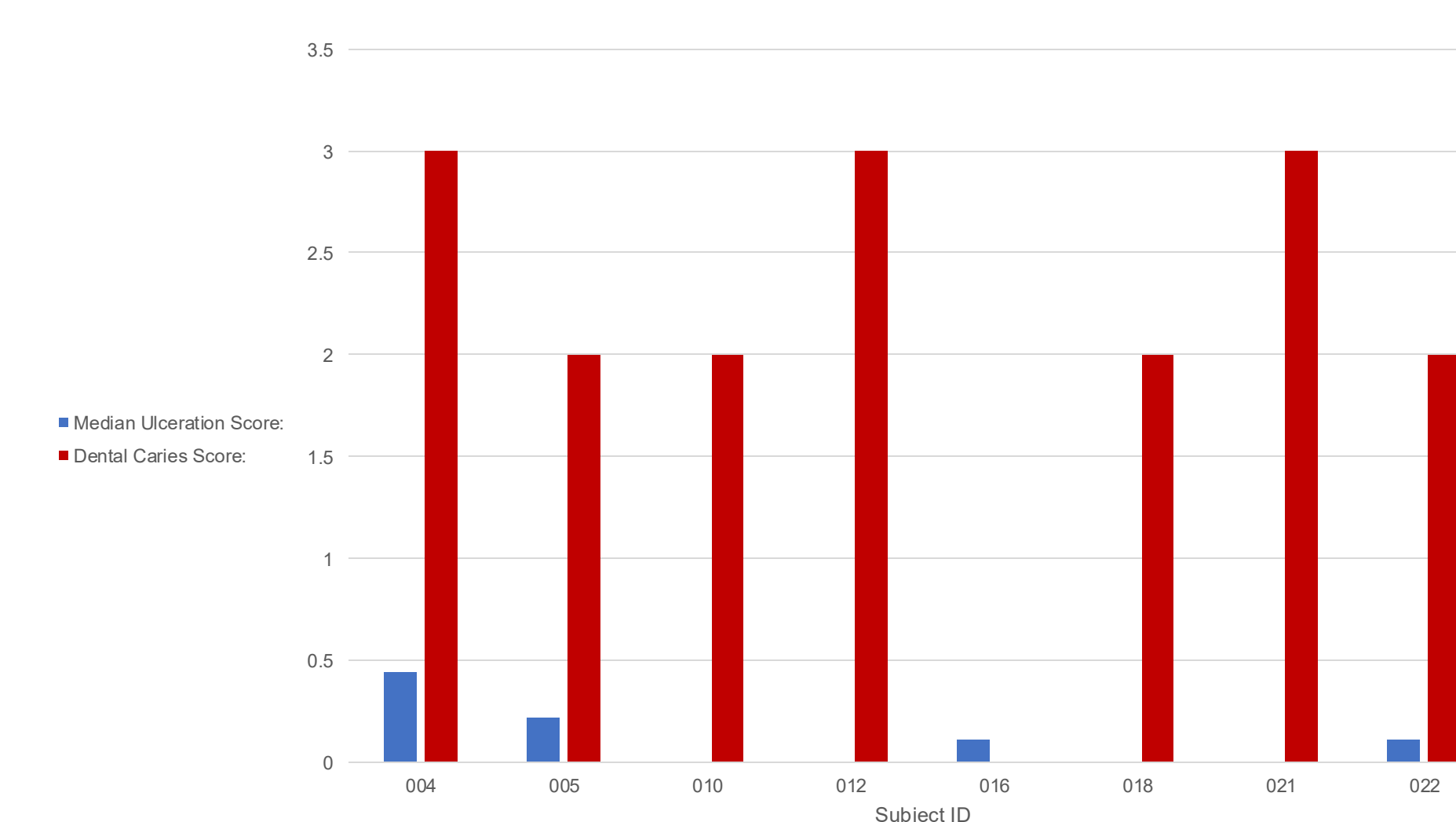
Data were collected from 22 subjects with a mean age of 17.1 ± 5.6 years.

77% of patients had a diagnosis of Fanconi Anemia (n=17), 18% with Dyskeratosis Congenital (n=4), and 5% with Shwachman-Diamond Syndrome (n=1).

Mean scores included OHI-S (0.93 ± 0.54), MGI (0.99 ± 0.59), and OMAS ($<0.04 \pm 0.10$). Patients had a mean caries score of 0.77 ± 1.2 , corresponding to 1-3 teeth with clinical caries. One outlier had non-restorable caries along with the highest OHI-S and MGI scores.



Results were consistent across subjects for median OHI-S and MGI. These indices assessed oral hygiene and baseline gingival inflammation at the time of the intraoral examination.



Subjects with positive results for median ulceration score and dental caries score are displayed. The majority of subjects (64%) had scores of 0 in these categories, indicating no clinical caries or ulcerations.

Conclusions

- IBMFS patients were recorded to have low-moderate gingival inflammation recorded.
- IBMFS Patient scores averaged good-to-fair oral hygiene
- Development-based recommendations from dental home should be followed..

Future Research Directions

This study serves as a pilot study for an ongoing partnership between the Cancer & Blood Diseases Institute Clinic and the Division for Pediatric Dentistry & Orthodontics at Cincinnati Children's Hospital Medical Center. Future studies aim to discover if salivary biomarkers can aid with early screening and diagnosis of gingival inflammation, dental caries, and pre-malignant lesions. Salivary samples are relatively easy to collect and have been shown to have diagnostic value in other applications. For IBMFS patients, early diagnosis and intervention can change the trajectory of their disease and overall health.

References

