## **Invited papers**

## Evidence and Clinical Improvement: Current Experiences with Dental Implants in Individuals with Rare Disorders

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Dental implants have been successfully used in totally and partially edentulous individuals for several decades. The long-term success and survival rates of implants, as well as the personal benefits in quality of life, are well described in multiple studies of large patient materials. In a meta-analysis of prospective, longitudinal studies with follow-ups of at least 5 years, the rate of implant loss was reported in 100% of the studies, whereas biological complications were mentioned in only 40% to 60% and technical complications in 60% to 80%.<sup>1</sup> Many would agree that there is strong evidence for the use of dental implants with regard to success and failure rates. However, many other relevant clinical questions remain unanswered and will fall into the category of clinical improvement rather than evidence-based research. For example, reports of treatment with dental implants in individuals with rare diseases are limited and in most cases restricted to anecdotal reports and case presentations.

In an era when the concept of evidence-based medicine and dentistry dominates the research scene, it is important to emphasize that this concept was originally meant to improve the quality of methods of research in the treatment of national ailments and common health complaints. High levels of evidence are what we must aim for whenever possible, but "no top-level evidence" does not equal "no evidence"! The lowest level of evidence is attributed to consensus statements, case presentations, and the experiences of specialists and authorities.<sup>2</sup> For many clinical issues, these methods are the only ones available and thus are the *best possible* in the endeavour to slowly increase our knowledge. One such area is treatment outcomes in rare diseases.

In Sweden, rare diseases are defined as disorders or injuries resulting in extensive handicaps and affecting no more than 100 individuals per 1 million inhabitants.<sup>3</sup> Dental and craniofacial development is affected in many rare diseases, since the mouth and teeth, with their complicated development and neurological functions, are among the most vulnerable to malformation and malfunction. Therefore, dentists can play a major role not only in treatment but also in diagnostics. The number of

# Table 1 Number of Syndromes That Match Certain Search Terms in the LDDB\*

Search term	No. of syndromes	
Face	1080	
Oral region	837	
Mouth	677	
Teeth	535	

\*London Dysmorphology Database, Oxford Medical Databases, Oxford University Press, version 2.2, 2000.

syndromes in the facial area that are listed in response to certain search terms in the London Dysmorphology Database (LDDB, Oxford Medical Databases, Oxford University Press, version 2.2, 2000) are presented in Table 1. For instance, the feature oligodontia—a condition that in many cases demands extensive multidisciplinary specialist treatment including prosthetic rehabilitation—occurs in 142 heritable syndromes.

When there are no references in the scientific literature, the first case report on treatment with a certain method in a specific syndrome will increase our knowledge by 100%. The challenges of the medical, biological, psychological, and technical difficulties associated with the different disorders make it particularly important to communicate any findings, favorable or unfavorable.

One of the questions most often asked at our oral disability center is whether it is possible to use dental implants in the oral rehabilitation of individuals with rare disorders. This has led to an effort to compile current experiences of treatment with dental implants in individuals with a diagnosis of a rare disease or syndrome. A search on PubMed in August 2005 for the terms rare disease and dental implant found 1 reference to a publication in a peer-reviewed journal, which was a case presentation on treatment with bone grafts and implants in a patient with systemic mastocytosis.<sup>4</sup> Down syndrome is not rare, but is the most common syndrome caused by a mutation. And although oligodontia is a common feature of this syndrome, Down syndrome has to date been the subject of only 1 case report on treatment with dental implants.<sup>5</sup> Ectodermal dysplasia is probably the heritable syndrome that is most well known to prosthodontists and 1 of the most well represented in the dental literature. Many case presentations have been made, but to date, only 1 prospective study on rehabilitation with dental implants in individuals with hypohidrotic ectodermal dysplasia has been published.<sup>6</sup> Two reports on treatment with dental implants in individuals with disabilities and chronic diseases were recently published in this journal: a prospective study on individuals with **Table 2** Publications on Treatment With Dental Implants

 in Rare Diseases and Syndromes

Rare disease or syndrome	Publications	
Cleidocranial dysplasia	Lombardas and Toothaker, 1997 <sup>9</sup> Petropoulos et al, 2004 <sup>10</sup>	
Ectodermal dysplasia	Guckes et al, 2002 <sup>6</sup>	
Epidermolysis bullosa	Penarrocha-Diago et al, 2000 <sup>11</sup>	
Erdheim-Chester disease	Brahim et al, 1992 <sup>12</sup>	
Gorlin syndrome (nevoid basal cellcarcinoma syndrome)	Markt, 2003 <sup>13</sup>	
Hypophosphatasia	Bergendal and Ljunggren, 2001 <sup>14</sup>	
Osteogenesis imperfecta	Ambjørnsen, 2002 <sup>15</sup> Lee and Ertel, 2003 <sup>16</sup>	
Papillon-Lefevre syndrome	Adbulwassie et al, 1996 <sup>17</sup> Ullbro et al, 2003 <sup>18</sup> Woo et al, 2003 <sup>19</sup>	
Systemic mastocytosis	Thor et al, 2005 <sup>4</sup>	
X-linked hypophosphataemic rickets	Bergendal and Ljunggren, 2001 <sup>14</sup>	

#### Table 3 The "Twin Sisters" of Science\*

#### Disease biology

The science of disease biology is the *hypothesis-driven* observation, identification, description, experimental investigation, and theoretical explanation of the phenomena associated with disease, *with the goal of preventing, treating, or eliminating it.* 

Anatomy, Physiology, Biochemistry, Genomics, Molecular biology, Pathology, Experimental design, Immunology, Laboratory management, etc.

#### Clinical practice

The science of clinical practice is the observation, identification, description, experimental investigation, and theoretical explanation of the phenomena associated with the *relief of the human burden of illness in daily clinical care for patients*.

Systems thinking, Informatics, Narrative research, Small groups, Psychology, Safety sciences, Epidemiology, Decision-making, Operations research, etc.

\*From Batalden P, MD, Director, Health Care Improvement and Leadership Development Center for the Evaluative Clinical Sciences, Dartmouth Medical School, Hanover, NH, USA.

neurologic disabilities<sup>7</sup> and a case series on special care patients.<sup>8</sup>

Information on the diagnoses of rare disorders in other sources such as published abstracts of posters and presentations at scientific meetings, workshops, and consensus conferences is presented in Table 2.<sup>9-19</sup> It adds enormously to the credibility of our profession to be able to report some experiences in the use of a proposed method of treatment to patients and families.

Dr Paul Batalden of the Institute for Healthcare Improvement (www.ihi.org) has visualized the "twin sisters" of science-the science of disease biology and the science of clinical practice (Table 3)-as being intertwined like the spirals of the DNA molecule, which underlines that it is not a question of either one or the other, but of both. Clinical improvement is often described in settings with low levels of evidence, and such research must ideally aim for higher levels. Nonetheless, some clinical results are best communicated as single case reports or consensus statements, which are valuable efforts in clinical (quality) improvement and which ought to be encouraged and published.<sup>20</sup> The medical journals Quality in Health Care<sup>21</sup> and the British Medical Journal<sup>22</sup> have made a commitment to publish papers on quality improvement. The new focus for this journalmanagement of patients' oral rehabilitative needspromises to give a voice to a broad spectrum of scientific and clinical issues.

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