

Survival Rates of Dental Implants in Patients with Papillon–Lefèvre Syndrome: A Systematic Review

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ABSTRACT

Aim: The present review aimed to summarize and evaluate the available literature regarding the survival rate and outcomes of dental implants in patients with Papillon–Lefèvre syndrome (PLS).

Materials and methods: An extensive search of the literature was conducted on PubMed, Scopus and Web of Science databases for all data published from January 1996 till April 2020 using a combination of the following keywords: 'Papillon Lefèvre Syndrome', 'prosthodontic rehabilitation' and 'dental implant' according to the PRISMA guidelines for the focused research question constructed using the PICO criteria. Clinical trials and observational studies on implant placement in PLS patients reported in English language were included in the study.

Results: A total of 10 studies (nine case reports and one case series) comprising 124 dental implants placed in 13 PLS patients were included. The follow-up period ranged from 4 months to 9 years. With regard to implant loading, 9 studies reported delayed loading, while one study did not provide any information regarding the nature of implant loading. The design of prosthodontic superstructure was either a removable or fixed prosthesis. Out of the 124 inserted implants, 20 (16%) were reported as failed. The overall survival rate was 84%.

Conclusion: The limited available evidence suggests that the survival rate of dental implants in patients with PLS is lower than that among healthy individuals. Nevertheless, no strict contraindication for implant-supported prosthesis seems to be justified in this group of patients. Further longitudinal studies with adequate follow-up periods are highly warranted.

Clinical significance: The prognosis of implant treatment for PLS patients has not yet been established. Dental practitioners should follow a careful approach in planning the dental implant treatment for this cohort of patients.

Keywords: Dental implants, Papillon–Lefèvre syndrome, Prosthodontic rehabilitation, Survival rate.

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BACKGROUND

Papillon–Lefèvre syndrome (PLS), first described in 1924,¹ is an autosomal recessive disorder affecting mainly 1–4-year-old children.² Although the etiology of PLS is not yet fully clear, PLS is caused by the mutation of the *cathepsin C (CTSC)* gene^{3,4} and has a higher prevalence in children born to consanguineous marriages.⁵ Globally, the prevalence of the disease is 1–4 persons per million,⁶ and both males and females of all races are equally affected.⁷

PLS is characterized by palmoplantar keratosis and severe periodontitis.^{8–11} Severe periodontitis can lead to the premature loss of deciduous teeth by the age of 4.^{11,12} Although gingival inflammation subsides soon after exfoliation of deciduous dentition, periodontitis re-occurs shortly after the eruption of the permanent teeth. Chronic periodontitis can lead to complete edentulism in children as young as 12 years.¹⁰ A combination of periodontal therapy,¹³ administration of drugs, such as antibiotics^{14,15} and retinoids^{11,16} along with the extraction of periodontally compromised teeth¹⁷ can be used in an attempt to impede the progress of periodontal disease and preserve the remaining teeth. However, the response to periodontal therapy is poor,^{10,18,19} and the subsequent loss of the remaining permanent teeth is mostly inevitable.²⁰ Moreover, the progression of periodontal disease would result in severe loss of the alveolar bone, creating prosthodontic complications.^{21–23}

The prosthodontic rehabilitation of children with PLS is quite challenging; it usually starts with removable partial dentures and

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shifts to removable complete dentures when the patient becomes fully edentulous.^{24,25} With the advancement of dental implants, the use of implant-supported overdenture (ISOVD)/fixed dental

prosthesis (ISFDP) in patients with PLS has also been reported.^{26,27} However, as stated above, several challenges are faced by the patients with PLS receiving prosthodontic rehabilitation. In addition to the psychological and physiological impact of edentulism,²⁸ the severely resorbed and atrophic alveolar ridge can undermine the rehabilitation of the patients with PLS.²⁹ The presence of an atrophic ridge covered with a thin mucosa can result in unstable dentures, oral discomfort, pain, and irritation.³⁰ Furthermore, inadequate bone can hamper the success of implant-retained dental prostheses and complicate the surgical intervention.³¹ It is also not yet evident whether patients with PLS have a greater risk for the development of peri-implantitis and resorption of the supporting bone around dental implants.

Several studies have reported the use of dental implants in patients with PLS; however, no such attempt has been made to systematically review the current literature on the outcomes of implant-supported prosthetic dental restorations in this group of patients. Hence, the objective of this study is to summarize and evaluate the available literature regarding the survival and outcomes of dental implants in patients with PLS.

MATERIALS AND METHODS

Focused Question

The present systematic review was conducted according to the Preferred Reporting Items for Systematic Review and Meta-Analyses (PRISMA) guidelines.³² A focused research question was constructed using the PICO (participants, intervention, control, and outcomes) criteria.³³ The focused question was: “What is the survival rate of dental implant therapy in patients with PLS?”

Search Strategy

An electronic search of databases (PubMed, Scopus, and Web of Science) was performed to identify all relevant articles using a combination of the following keywords: “Papillon-Lefèvre syndrome,” “prosthodontic rehabilitation,” and “dental implant” for all data published from January 1996 to April 2020. Also, a manual search of the reference lists of the retrieved articles was conducted to identify further studies. Two independent reviewers (SAA and HMA) screened titles and abstracts of all studies retrieved

by the research strategy, and irrelevant studies were excluded. Full articles were sought for all potential studies and reviewed by the two reviewers for inclusion.

Eligibility Criteria

Predefined eligibility criteria were used to screen all identified studies. Studies were included if they fulfilled the following criteria:

- Human subjects.
- All types of clinical trials and observational studies.
- Studies reporting on implant placement in patients with PLS.
- Studies reporting on survival or failure of dental implants in patients with PLS.
- Studies in the English language.

Experimental studies, animal studies, review articles, and letters to editors were excluded.

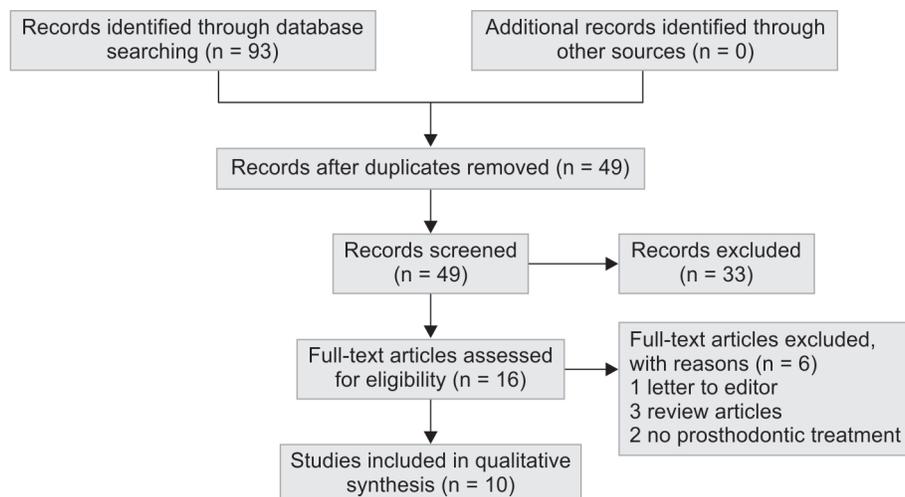
Data Extraction

The following data were extracted using a special data collection form: authors and year of study, study design, country of the study, number of patients, patient’s gender, patient’s age at the start of implant therapy, number of dental implants, implant dimensions (diameter and height), implant type, adjunct procedures, type of implant loading, type of prosthodontic superstructure, reported follow-up time after the implant treatment, and main outcomes and conclusion.

REVIEW RESULTS

Study Selection

The initial online search yielded a total of 93 publications, 44 of which were duplicates and were thus excluded. After screening the titles and abstracts, 33 studies were found to be irrelevant and were therefore removed. Full-texts of the remaining 16 studies were sought and thoroughly read by the two authors (SA Al-Maweri and HM Al-Shamiri). Of these, six publications were excluded because they did not meet the eligibility criteria. Ultimately, 10 studies were included in the systematic review. The results of the search strategy are illustrated in [flowchart 1](#).



Flowchart 1: Flowchart of methodology

Table 1: General characteristics of the included studies and treated patients

Study	Type of study	Country	No. of treated patients (n)	Patient's gender		Patient's age at the start of implant therapy (years)
Ullbro et al. ³⁷	Case report	Saudi Arabia	1	F		25
Woo et al. ³⁴	Case report	USA	1	M		14
Toygar et al. ³⁵	Case report	Turkey	1	F		18
Etöz et al. ²⁶	Case report	Turkey	1	F		34
Ahmadian et al. ³⁸	Case report	Iran	1	F		21
Senel et al. ²⁹	Case report	Turkey	1	M		18
Al Farraj AlDosari ²⁷	Case report	Saudi Arabia	1	F		19
Nickles et al. ⁴⁰	Case series (retrospective study)	Germany	4	P1	F	20
				P2	F	20
				P3	F	Max: 34 Man: 18
				P4	F	Max: 21 Man: 19
Rai et al. ³⁶	Case report	India	1	M		26
Kinaia et al. ³⁹	Case report	USA	1	M		21

M, male; F, female; P, patient; Max, maxilla; Man, Mandible

General Characteristics of the included Studies and Treated Patients

A total of 10 studies (9 case reports and 1 case series) comprising 13 patients with PLS with 124 implants were included (Table 1).^{26,27,29,34–40} Three studies were conducted in Turkey,^{26,29,35} two in the USA,^{34,39} two in Saudi Arabia,^{27,37} one in Germany,⁴⁰ one in Iran,³⁸ and one in India.³⁶ Out of the 13 treated patients, 9 were females and 4 were males. The age of patients at the start of the implant treatment ranged between 14 and 34 years. Table 1 presents the former findings.

Variables Related to Implant Treatment

Table 2 outlines variables related to implant treatment. A total of 124 dental implants were inserted in 13 patients with PLS. Out of 124 implants, 61 were placed in the maxilla and 63 in the mandible. The number of inserted implants varied from case to case and ranged from 2 to 20 implants. The diameter of the implants was reported in five studies^{26,27,34–36} with a range from 3 to 5 mm. The height of the implants was mentioned in six studies^{26,27,34–37} and ranged from 6 to 13 mm. In four studies, adjunct procedures, such as bone augmentation and bone grafting, were used to optimize the ridge site for implant placement.^{27,35,38,39} With regard to the type of implant loading, nine studies reported delayed loading ranged from 3 to 15 months^{26,27,29,34–39}, and one study did not provide any information regarding the nature of implant loading.⁴⁰ The design of the prosthodontic superstructure was either a removable or fixed prosthesis supported by dental implants. The follow-up time after implant insertion ranged from 4 months to 4.5 years in 12 cases. In one patient, the follow-up period extended over 9 years.

The Main Outcomes

The overall survival rate of dental implants was 84%. Out of 124 inserted implants, 20 implants (16%) were reported as failed. Around two-thirds of the failed implants ($n = 14$) were initially placed in the maxilla and six (30%) were in the mandible (Table 2).

Of the failed 20 implants, the majority (19 implants) were reported in one study by Nickles et al.,⁴⁰ who retrospectively evaluated the outcomes of implant therapy in 4 patients with PLS who received 54 dental implants. Three patients experienced

varying degrees of peri-implantitis, and peri-implant mucositis was detected 1 year after the placement of 10 implants in the fourth patient. The loss of implants was recorded among two out of the former four patients with PLS. In the first case, four out of six maxillary implants failed after 2 years of observation. In the second case, failure affected 15 out of 20 inserted implants (9 in the maxilla and 6 in the mandible) (Table 2). The authors attributed the unfavorable results among this group of patients with PLS to lack of compliance toward recall visits and oral hygiene measures.⁴⁰ On the other hand, the other 9 selected case reports indicated successful outcomes of 70 dental implants in 9 patients with PLS at the follow-up time (Table 2).^{26,27,29,34–39} Success was in terms of implant osseointegration, preservation of supporting bone, oral function, and patient satisfaction. However, in the case reported by Senel et al.,²⁹ one maxillary implant failed at 1 month after implant surgery. In this case, six implants were initially inserted in the maxilla and six in the mandible. The failed maxillary implant was then replaced and the replacement implant survived over the follow-up period (Table 2).

DISCUSSION

The prosthodontic rehabilitation of the patients with PLS can be considered a prosthodontic dilemma. Although the conventional prosthodontic rehabilitation with removable dentures provides a relatively simple restorative solution for the PLS patient, achieving adequate oral comfort and function is probably an unattainable objective with this treatment option for the patients with PLS.^{26,34,37}

Moreover, treatment with tooth-supported fixed partial dentures is mostly contraindicated in the patients with PLS due to young age and lack of abutment teeth that can retain and support a fixed dental prosthesis. This is coupled with significant psychological, social, esthetic, and functional damage affecting the PLS patient that has become completely or partially edentulous at an early stage of life. Under the aforementioned circumstances, the use of dental implants to retain and support a dental prosthesis becomes tempting for both clinicians and patients with PLS. The well-known superior treatment outcomes with implant-supported prostheses lead to such a treatment decision.⁴¹ However, no solid evidence is yet available to guide the decision-making process in

Table 2: Variables related to the implant treatment and final reported outcome

Study	Treated patients (n)	Number of implants (n)	Implant dimensions (mm)		Implant type	Adjunct procedures	Type of implant loading	Prosthesis/ prosthodontic superstructure		Follow-up after implant treatment	Outcome and conclusion
			Diameter	Height				Max	Man		
Ullbro et al. ³⁷	1	Max: 0 Man: 5	NR	10	ad modum Brånemark (Nobel Biocare, Gothenburg, Sweden)	None	Delayed (3 months)	CD	ISFDP	4.5 years	4.5 years after the implant installation, the treatment was clinically and radiographically successful
Woo et al. ³⁴	1	Max: 0 Man: 2	4	13	Branemark system titanium implants (Nobel Biocare, Goteborg, Sweden)	None	Delayed (4 months)	CD	ISOVD	1 year	Successful osseointegration and preservation of alveolar bone 1 year after implant placement and the continual wearing of a functional dental prosthesis
Toygar et al. ³⁵	1	Max: 2* Man: 0	3.7	13	Tapered Screw-Vent, Zimmer Dental, Carlsbad, CA	Alveolar bone augmentation	Delayed (4 months)	ISFDP	RPD	4 months	A multidisciplinary approach with advanced periodontal surgery, orthodontic and prosthetic treatment, and implant therapy may be an appropriate treatment modality for dental rehabilitation in patients with PLS
Etöz et al. ²⁶	1	Max: 0 Man: 2	4.1	6	ITI Dental Implant System, Straumann AG, Waldenburg, Switzerland	None	Delayed (3 months)	CD	ISOVD	1 year	After 1 year of follow-up period, the clinical and radiological conditions of the osseointegrated implants and lower denture were acceptable, and there was no sign of infection and/or unexpected bone loss around the implants
Ahmadian et al. ³⁸	1	Max: 8 Man: 8	NR	NR	ITI SLA implants (Institute Straumann AG, Waldenburg, Switzerland) with regular platforms	-Orthognathic surgery with set-back of mandible -Inferior alveolar nerve repositioning -Bilateral sinus grafts -Vestibuloplasty procedure	Delayed Max: 6 months Man: 15 months	ISFDP	ISFDP	4 years	At the 4-year follow-up appointment, the patient presented significant improvements in oral function and psychosocial activities and no prosthetic complications

Contd....

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Study	Treated patients (n)	Number of implants (n)	Implant dimensions (mm)		Implant type	Adjunct procedures	Type of implant loading	Prosthesis/ prosthodontic superstructure		Follow-up after implant treatment	Outcome and conclusion
			Diameter	Height				Max	Man		
Senel et al. ²⁹	1	Max: 6* Man: 6	NR	NR	ITI Dental Implant System; Institute Straumann AG, Waldenburg, Switzerland	None	Delayed (6 months)	ISOVD	ISFDP	3 years	At 3 years follow-up, all implants were clinically stable and no pain or infection was found
Al Farraj AlDosari ²⁷	1	*1 month after implant surgery a maxillary implant was mobile due to lack of osseointegration The failed implant was then replaced. Max: 8 Man: 6	3.5/4/5	9/11/13	OsseoSpeed™, Astratech Dental Implant System, Mölndal, Sweden	Bone grafting in the premolar and molar regions	Delayed (4 months)	ISFDP	ISFDP	1 year	A one-year follow-up of the case showed a functionally and esthetically stable dental implant with no signs of infection or bone loss
Nickles et al. ⁴⁰	4	P1 Max: 6 Man: 4	NR	NR	NR	NR	NR	NR	NR	P1 4 years	Some of the implants showed peri-implantitis 4 years after placement
		P2 Max: 6 Man: 4								P2 1 year	Peri-implant mucositis was found one year after implant placement
		P3 Max: 6 Man: 8								P3 2 years	4 maxillary implants failed
		P4 Max: 10 Man: 10								P4 Max: 7 years Man: 9 years	The two survived maxillary implants presented with advanced peri-implantitis
Rai et al. ³⁶	1	Max: 0 Man: 2	3/3.3	10/11.5	NR	None	Delayed (6 months)	CD	ISOVD	1 year	Successful 1-year follow-up of implant osseointegration and alveolar bone preservation
Kinaia et al. ³⁹	1	Max: 9 Man: 6	NR	NR	Legacy 3, Implant Direct	Bone augmentation using autogenous cranium parietal bone graft (in both jaws)	Delayed (3 months)	ISFDP	ISFDP	1 year	Well-osseointegrated implants with good function and esthetic smile. The patient was satisfied with the treatment outcomes

Max, maxilla; Man, mandible; NR, not reported; P, patient; CD, complete denture; RPD, removable partial denture; ISOVD, implant-supported overdenture; ISFDP, implant-supported fixed dental prosthesis

the management of patients with PLS. To date, the prognosis of dental implant therapy in patients with PLS cannot be predicted. The current review can be considered the first to evaluate the available evidence regarding the survival rates of dental implants in patients with PLS.

The analysis of the extracted data shows that nine of the selected studies illustrated positive outcomes at the end of the follow-up period of the implant treatment for the patients with PLS. On the contrary, the single included retrospective study presented poor survival rates of dental implants (Table 2). Out of 46 inserted implants, 20 showed failure in 3 patients with PLS.^{29,40} Moreover, in 10 patients with PLS^{26,27,34-40} 78 inserted dental implants remained in function at the time of observation (Table 2). The big picture suggests that the dental implants have a reasonable chance to survive in the patients with PLS (84% survival rate). However, the results of the present review should be interpreted with caution considering the apparent limitations, discussed in the following sections.

It can be noted that the selected studies can provide only a low level of evidence as they were nine case reports and one retrospective study. Furthermore, these studies were heterogeneous in terms of patient's age at the start of implant therapy, the number of inserted implants, the implant dimensions, the implant type, the use of adjunct procedures, the design of prosthodontic superstructure, and the duration of follow-up after the implant treatment (Tables 1 and 2).^{26,27,29,34-40} Though many factors can be involved in the failure of dental implants,⁴²⁻⁴³ the heterogeneity and the design of the selected studies limit the ability to have a profound understanding of the factors that may enhance or risk the survival of dental implants in the patients with PLS. Nevertheless, several remarks can be expressed in this respect.

An adequate follow-up period is crucial to establish the 5- and 10-year survival rate of the dental implants. The present literature indicates that the 5- and 10-year survival rate of dental implants among the general population is up to 98 and 96%, respectively.⁴⁴⁻⁴⁶ In this review, most of the included studies showed encouraging results for the dental implant treatment among the patients with PLS, as 84% of the inserted implants survived at the time of follow-up. This figure is lower than that observed among the general population for the survival rate of dental implants. Moreover, this finding is mostly based on short-term observations of between 4 months and 4.5 years. Only one patient with PLS was followed-up for 9 years after implant therapy and exhibited a high failure rate among the 20 placed dental implants (75%).⁴⁰ It seems that dental implants provided for patients with PLS have a good short-term survival rate but in the long-term, the risk of failure might be alarming. Nickles et al.⁴⁰ attributed the high failure rate of dental implants in the above-mentioned PLS patient to poor compliance with oral hygiene measures and maintenance therapy. The authors indicated that even the five survived implants in this patient with PLS exhibited an advanced level of peri-implantitis. They concluded that the risk of peri-implantitis and implant loss can be quite high in the patient with PLS, who lacks the motivation to maintain an adequate level of oral health.⁴⁰ On the other hand, nine included case report studies reported very cooperative and compliant patients with regular follow-up, which may explain the high survival rates of implants in these patients (only 1 out of 70 inserted implants failed). Given this and the fact that PLS is usually characterized by the development of severe periodontitis, post-treatment recall, and effective oral hygiene maintenance may be major factors that can

impact the survival rate of implants placed in patients with PLS. Whether progression of peri-implantitis is a character of the PLS in the presence of dental implants vs the advancement of periodontitis in the presence of teeth, is not evident.

One recent systematic review on post-loading implant loss found a significantly higher rate of implant loss in the maxilla compared to the mandible.⁴⁷ In this review, 70% of the failed implants ($n = 14$) were in the maxilla vs six implants (30%) failed in the mandible (Table 2). The risk of implant failure may be higher in the maxillary arches of the patients with PLS compared to mandibular arches.

Bone availability is a determining factor for the success of dental implants.^{42,43} This issue is even more critical in patients with PLS given the severe bone loss in these patients. Among the included studies, bone augmentation was performed in four patients with PLS before implant insertion.^{27,35,38,39} Interestingly, these reported cases showed positive outcomes reflected by good implant stability, well-functioning prosthesis, and lack of any sign of infection or bone loss at follow-up. It appears that adjunctive procedures, such as bone augmentation, can be used when needed with patients with PLS to optimize implant sites and facilitate implant placement.

In the studies reviewed for this article, implants have been used to retain and support both removable and fixed prosthetic restorations in patients with PLS, and no apparent differences can be identified in the efficacy of ISOVD and ISFDP in the light of this systematic review. However, it is known that the design of ISFDP requires a greater number of dental implants, a wider range of prosthetic and surgical procedures, a higher standard of oral hygiene maintenance, and a more intensive periodontal recall program compared to ISOVD. A study by Nickles et al.⁴⁰ strongly suggests that poor patient compliance can adversely affect the outcome of implant therapy in patients with PLS and, if combined with a complex design of ISFDP, it can have a deleterious effect on the outcome of treatment.⁴⁰ Furthermore, the burden of the failure of an implant on the design and the prognosis of an ISFDP may exceed that with ISOVD. It is perhaps more prudent to opt for ISOVDs in the rehabilitation of patients with PLS to minimize the risk of post-treatment complications.

It is clear that the nine included case reports utilized a delayed protocol of implant loading. This type of implant loading might contribute to the survival of dental implants in the patients with PLS as failure affected 1 out of 70 inserted implants (Table 2). In the retrospective study of Nickles et al.,⁴⁰ no information was provided with regard to the type of implant loading. What is the outcome of immediate loading of implants in the patients with PLS is not known? Seemingly, the delayed implant loading is the suggested protocol of implant loading in the patients with PLS till solid evidence is available to guide treatment decision making in this respect.

As previously elaborated, most patients with PLS eventually lose all their teeth before adulthood. An important question to be raised here is about the right age to insert dental implants in the jawbones of patients with PLS. The literature indicates that placement of dental implants in young patients before maturity has several clinical implications that may negatively affect the outcome of the implant treatment.^{48,49} Implants act like ankylosed teeth if they are placed in growing jaws, and this can result in esthetic, functional, and periodontal complications once the growth of the jaws has stopped.^{48,49} Woo et al.,³⁴ reported placement of dental implants in a growing PLS patient. However,

the authors reported a 1-year follow-up period for this patient, which is a very short time to reveal any possible complications associated with placement of dental implants at a young age. Based on the former arguments, it would be more advisable to postpone the implant treatment for patients with PLS till the attainment of maximum jaw growth.

Although the present systematic review provided an important insight into the survival rates of dental implants in PLS patients, it has several limitations that should be taken into consideration. The main limitation is the fact that the evidence obtained is based on case reports. Another important limitation is the marked heterogeneity among the included studies with respect to implants parameters and time of loading after implant placement. Additionally, the short follow-up time in most of the included studies is an obvious limitation.

Overall, this review has underlined a big gap in the current knowledge about the outcomes of dental implant treatment in patients with PLS. A piece of evidence-based knowledge is lacking. Case reports studies of short follow-up time constitute the major bulk of the available literature regarding the provision of dental implants for patients with PLS. The need for future well-designed prospective cohort studies or randomized clinical trials is apparent for the sake of a more in-depth understanding of the outcomes of the implant therapy in patients with PLS and the factors that affect the survival rate of dental implants and long-term treatment success in this group of patients. Due to the nature of PLS, research collaboration on an international level is highly recommended to document and monitor the patients with PLS who are subjected to the implant treatment. A possible role for the World Health Organization (WHO) may be critical in this aspect.

Till the time robust evidence is present, the authors recommend a careful approach in planning the dental implant treatment for the PLS patient. This includes an adult PLS patient, a minimum number of dental implants, a delayed-type of implant loading, a prosthesis of removable design, a meticulous level of oral hygiene, and regular recall maintenance visits. From an ethical standing, the patients with PLS should be informed that their implants have a fair chance to survive but the prognosis of the implant treatment for this cohort of patients has not yet been established.

CONCLUSION

It appears that the short-term survival rate and outcomes of the dental implant treatment in the patients with PLS are encouraging. However, the long-term survival rate of dental implants in patients with PLS is not evident. Well-designed future studies with adequate follow-up periods are required to bridge the gap in our current knowledge about the survival of dental implants and factors affecting their success in this group of patients.

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