

# Dental treatment of patients with prune belly syndrome

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## INTRODUCTION



**Prune belly syndrome (PBS),** also known as **Eagle-Barrett syndrome (EGBRS),** is a rare congenital disease characterized by deficiency or absence of abdominal wall muscles, urological abnormalities, and bilateral cryptorchidism.

## KEY FINDINGS

Prune belly syndrome can be classified based on antenatal and postnatal features into three categories.

**Category 1 PBS** includes patients with severe renal dysfunction and pulmonary hypoplasia. It has almost 100% mortality rate.

**Category 2 PBS** patients display the classic triad features with varying degrees of renal dysplasia.

There is a large variation in severity of PBS, some patients may need an early dialysis.

**Category 3** patients have a normal renal function and mild phenotypic features of PBS.

Dental changes such as enamel hypoplasia predispose to caries. Some PBS patients present with a cleft lip or gingival fibromatosis. Of all the medical issues of PBS, renal impairment has the largest effect on the patient's health status.

The majority (90 %) of patients with chronic renal disease suffer from oral symptoms from their syndrome and their medications. They include but are not limited to gingival bleeding, gingival hyperplasia, pulp obliteration, delay or alteration of tooth eruption, osteoporosis and bone loss, infections (most frequently candidiasis) and xerostomia .

Category	Clinical features	Adjustments							
		Blood Pressure	Void Prior to treatment	Consult Primary Care MD or Nephrologist	Reduction in prescribed medication	Reduction or elimination of anesthetic	Evaluation of xerostomia and possible solution	Pre-med	Tx on non dialysis day
Category 3	Mild phenotypic features of PBS	✓	✓	PCP					
Category 2	Moderate phenotypic feature PBS & mild renal impairment	✓	✓	PCP or nephrologist					
	Most all phenotypic feature of PBS & moderate renal impairment	✓	✓	Nephrologist	Consider	Consider	Stannous fluoride or change of xerostomia meds.		
	All phenotypic features of PBS & severe renal impairment	✓	✓	Nephrologist	✓	✓	✓	possible	✓
Category 1	Severe renal impairment and pulmonary hypoplasia	✓	✓	Nephrologist	✓	✓	✓	✓	✓
Category 1	Close to 100% mortality				Hospital based dentistry.				

## METHODS

We reviewed research articles published during the time from 1965 to 2021 using four search engines PubMed, Scopus, and Google Scholar.

Individually or in combinations, we used keywords prune belly, prune belly syndrome, PBS, Eagle-Barrett, dental manifestation, clinical manifestation, psychological aspects.

The search was run with no language restrictions. We obtained 522 articles on PBS and 11 articles on EGBRS. Interest in PBS started to increase in late eighties and has continued. The majority of these articles dealt with diagnosis and treatment.

We focused on information related to oral health and dental care.

## RESULTS AND DISCUSSION

**Dental Treatment:** Preparing for a dental treatment of a patient with PBS requires a consultation with the patient's physician to obtain an information about the patient's health and about a degree of impairment of various organs, especially the kidney.

Questions to be included in the conversation with the physician are : (1) what category of PBS does the patient fall into, (2) what organs have been affected and to what degree, (3) what medications is the patient correctly taking, (4) does the physician recommend premedication with antibiotics, (5) should the dosage of administered drugs be adjusted to decreased function of kidney (6) are there any other modifications to dental care that the physician can recommend.

**Tele dentistry:** As with any patient who has impaired organ function and may be immunocompromised either due to presented medical issues or due to prescribed medications, a limited time in the dental clinic is advisable.

You can meet the patient without facial covering which is less frightening for young children.

Care for patients with PBS may have additional challenges for not only the dental provider but also for the patient and possible caregiver. One study evaluated the health-related quality of life for children with PBS and their caregivers and found a lower overall health-related quality of life (HRQoL) scores. PBS patients had HRQoL scores comparable to those of children with cerebral palsy.

## CONCLUSIONS AND RECOMMENDATIONS

Many patients with PBS need a special dental care because compromised renal and respiratory functions negatively affect their oral and dental health.

One obstacle with regards to dental care for these patients is a difficulty in finding a dentist who would fully understand variations of features and medical history of patients with PBS and who would be willing to provide compassionate and safe care to prune belly patients.

We believe that providing information to dental practitioners will improve their understanding of PBS, will help them to better treat these patients, and will encourage them to welcome patients with PBS into their practice.

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*References available upon request*