CASE REPORT
The 3 year old girl presented at our clinic in 2012 with no history of diseases or abnormal development except for premature loss of her deciduous front teeth in the upper and lower jaw. At the age of 2 1/2 the first loose tooth was noticed and shed half a year later. Until the age of 4 she had lost central and lateral deciduous incisors of the upper and lower jaw. While under orthodontic surveillance her lower deciduous canines loosened and shed within a year (Fig.1-3).

INTRODUCTION
We present a case of odontohypophosphatasia (odonto-HPP), a rare congenital disease characterized by a genetic deficiency of tissue-nonspecific alkaline phosphatase due to ALPL-gene mutation. Clinical features of HPP are premature loss of deciduous teeth without prior root resorption, deformities, and discoloration of teeth as well as the susceptibility to caries [1].

ANAMNESIS
Our patient was the youngest child in a family of two children. Pregnancy was without complications. Parents, siblings and grandparents were healthy and did not show any signs of unusual tooth loss. No bony deformation was detected. Due to the absence of adequate trauma, periodontitis or excessive load that would have been plausible to explain primary tooth loss a genetic background was suspected.

CHARACTERISTICS AND PATHOPHYSIOLOGY
Currently seven forms of ALPL-gene mutation presenting as hypophosphatasia are differentiated by means of disease onset (prenatal lethal, prenatal benign, early infantile, infantile juvenile, adult, odonto-HPP and pseudo-hypophosphatasia). Time of onset correlates with the impact on bone metabolism, odonto-HPP being the mildest form [1]. Alkaline phosphatase catalyzes formation of matrix for bones and cementum, and its reduction results in hypoplasia and defective mineralization of alveolar bone and cementum. Dental tissues are highly sensitive to phosphate metabolism dysregulation. Therefore it is hypothesized that milder forms of ALPL-gene mutation have an impact on dentition only [2].

CONCLUSION
Odonto-HPP is an inherited disorder of mineral metabolism that should be interdisciplinarily recognized by dental professionals as a possible cause of premature loss of deciduous teeth without prior root resorption. A therapy has not been introduced yet. For genotype-phenotype relations have not been defined clearly it is especially important to identify milder forms of mutations for genetic counseling of the parents.

REFERENCES