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# DENTAL MANIFESTATIONS OF HEREDITARY HYPOPHOSPHATEMIC RICKETS CLINICAL,RADIOGRAPHIC AND BIOCHEMICAL STUDY.

The X liniked hypophgosphatemia VDRR is the most common type of rickets. In this study ten children patients with different age and sex and clinically manifested VDRR were selected. Clinical, radiographic and chemical investigations were performed. From the result it was evident that the most common dental manifestations were gingival abscesses and fisula. Also, enamel hypoplasia, loosness of teeth and early shedding of the Decidous teeth.

## REVIEW OF LITERATURE:

Parathyroid hormone and vitamin D are the principal hormone products of vertebtrate evolution that regulate extracellular phosphorus and calcium homostasis(1). Vitamin D acts on small intestine to enhance absorption of both calcium and phosphate. Parathyroid hormone acts on the kidneys to conserve calcium and reject phosphate anion in glomerular filtrate(4). Inadequate calcium or vitamin D in diet or insufficient exposure to sunlight or both leading to rickets of alimentary origin and can be easily treated by supplying calcium and phosphate in diet and by administering an adequate amount of vitamin D(3).

There is another form of rickets which did not respond to vitamin D therapy. This condition was first described by Albright et al.1937(5) as vitamin D resistant rickets (V.D.R.R.) or a refractory condition. It is called also, hereditary hypophosphatemic rickets or rickets of renal origin(2). Hypophosphatemic trait is inherited through an X linked dominant gene where the abnormal mutant alley appears on the X chromosome(15,16).

The prime features of this disorder are familial occurence, lowered serum phosphate level associated with decreased renal reabsorption of phosphate and lack of response to physiological doses of vitamin D(5).

The clinical manifestations of the disease often appear in the second year of life, at the time affected children begin to walk. The children are of short stature and often have significant bowing of the legs. The extra-oral radiographic evidence revealed an increase anteroposterior diameter of the skull rachitic rosary and enlargement of wrists.

Dental alterations are the first observed sign of VDRR as spontaneous gingival and periapical abscesses without concurrent evidence of caries or root fractures. Due to the abnormalities in the dentin formation that evident in the histopathologic examination especially in the region of pulpal horns that reveals tubular defects of the dentin and elongated into pulp hornes under incisal edges(8,9).

In radiographic examination of the dental structures often reveals enlarged pulp chambers and pulpal horns of primary dentition, extending to the dentinoenamel junction and extensive osseous lesions of rarifaction which involve the entire root and suggesting a periodontal in origin(10). Many outhers(11-14) reported that there are many abnormalities affecting both deciduous and permanent teeth as looseness, large root canals and pulp chambers, enamel hypoplasia, attrition and loss of lamina dura.

The phyophosphaturia usually persists after administration of vitamin D and is probably due to an inborn error or phosphate homeostasis, manifested by a selective defect in secondary active transport of phosphate in the brush border membrane of the proximal nephron(17). The abnormal low inorganic serum phosphate, normal serum calcium levels and elevated serum alkaline phosphatase are common biochemical characteristic of this disorder(11).

## AIM OF THE STUDY:

The purpose of this study is to reveal the dental abnormalities in heriditary hypophosphatemic rickets or vitamin D resistant ricket (VDRR) by clinical, radiographic and biochemical examination.

## MATERIAL AND METHODS:

From in and out children hospital, Cairo University, ten children patients with age range (3.5 to 12 years) with history of heriditaryin- origin Clinically manifested VDRR were selected. They had,received dose therapy of 600,000 I.U. of vitamin D for three injections with no improvement as noted from radiographic findings and laboratory data of serum calcium and phosphorus and alkaline

البحث العلمى الحادى عشر

# الظواهر السنية فى مرض الكساح الموروث المقاوم لفيتامين (د)
## دراسة إكلينيكية – معملية لكمياء الدم وشعاعية
## DENTAL MANIFESTATIONS OF HERIDITARY HYPOPHOSPHATEMIC RICKETS, CLINICAL, BIOCHEMICAL AND RADIOGRAPHIC STUDY


الملخص العربى

إن الكساح المقاوم لفيتامين (د) هو مرض آيضى يحدث فى الطفوله ، وقد تعرفنا عليه اكلينيكيا منذ ثلاثين عاما، وقد اتضح انه يقاوم الجرعات المعتاده من فيتامين (د) وأنه لا يشفى الا بإستخدام جرعات كبيرة جدا منه وقد تم فحص ثلاثين مريضا بالكساح الظاهر اكلينيكيا وتراوحت أعمارهم من ٥ر٣ – ١٤ سنه ، وقد اختيروا من مستشفى أبوالريش للأطفال ، وقد تم فحص دقيق اكلينيكى للمرضى ، وأخذ الأشعة اللازمة لكل منطقة فى الفم ، وكذلك الرأس ، وأيضا تم عمل الفحوص الكيميائية الحيوية لتقدير مستوى الكالسوم وفسفور الدم والانزيم الفوسفاتيزم القلوى.

وقد كانت نتائج البحث تدل على :-

١) أن تقوص السيقان وإلتواء العمود الفقرى لا يحدث الا عند الجلوس أو المشى ، وكذلك عدم وضوح النتائج المعملية الخاصة بهذا المرض ، فى البداية نتيجة المدى المنخفض للترشيح المخفض فى الطفولة وهو الذى يمنع ظهور الفوسفات فى البول وبالتالى يمنع انخفاض للفوسفات فى الدم كعلامة مميزة لهذا المرض .

٢) ولقد وجد أن طبيب الاسنان هو أول مكتشف للأعراض المبكرة للمرض لأن هذا المرض عادة يبدأ بأعراض فموية وسنية ، وهذا يؤكد دوره فى ارشاد المريض الى العلاج الطبى السليم وبالتالى تجنب العواقب الخطيره للمرض.

٣) الأعراض السيئة الشديده فى المرض أمكن مشاهدتها بقليل من الأعراض العامة بواسطة الأشعة . والفحوص المعملية الدم ولذلك أمكن التوصل للتشخيص السليم للمرض .