A STUDY OF RICKETS — MORBIDITY AND AETIOLOGY OF A ‘LOW PROFILE’ DISORDER

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SUMMARY

In a prospective study undertaken at the Paeds 'A' Unit Hayat Shaheed Teaching Hospital Peshawar, a total of one hundred and sixty eight (168) cases of Rickets were registered over one year period. After clinical evaluation and investigations one hundred and twenty four cases (73.8%) out of the total were attributed to Vit D Deficiency. The remaining forty four cases, (26.2%) of Rickets in decreasing order of frequency were due to renal tubular disorders of phosphate absorption (Familial Rickets), malabsorptive states, chronic renal disease, hepatobiliary disorders, use of anticonvulsant drugs and associated with rare-disorders like renal tubular acidosis, de Toni-Fanconi, Di George syndromes. In the Vitamin D Deficiency group, the male infants and children (62%) predominated the female children (38%) whose mean age of presentation (23 months) was later than the male group (14 months). The commonest clinical presentation was in the form of recurrent lower respiratory infection(LRTI) (48%). In this group 60% patient had wheeze in addition to LRTI. Other patterns of clinical presentation were delayed motor development (28%), due to generalized hypotonia, diarrhoea and abdominal distention 22%, non febrile seizures 10%, breath holding attacks, and tetany in less than 10%. Seventy percent of the patients had one or more skeletal changes suggestive of Rickets on initial presentation but not noticed by the parents. In the Vit D. Deficiency group, the largest group of patients, approx. 47% belonged to the inner city multistory dwellings of old Peshawar while the rest came from the rural districts belonging to both Northern and Southern regions of NWFP. In this group the two major causes of Vit. D deficiency were inadequate dietary intake of Vit D, compounded by insufficient exposure to available sun light. Living in poorly sunlit housing, as well as environmental pollution from traffic, forming thick smoke and dust screen blocking off the ultraviolet light (essential for Vit D synthesis in the skin) appeared to be the main factor responsible for Vit D deficiency. Cultural practices of keeping the babies indoors and cladding them in heavy clothing as well as low Vit. D status of the pregnant and lactating mothers due to “Purdah” observation may be important in contributing to the aetiology of rickets, in the remaining subjects belonging to the rural community.

INTRODUCTION

Rickets is a metabolic disorder of growth affecting infants and young children. It particularly effects the bones, muscles and some time the nervous system. It commonly results from deficiency of Vit D (Cholecalciferol) and occasionally that of calcium and phosphate. The main sources of Vit D are its synthesis in the skin from 7-Dehydrocholesterol under the effect of ultraviolet light of the sun, intake in the
form of naturally occurring foods, Vit D fortified diets and dietary supplementation with medicinal preparations. Among these the most important source is the Vit D which is synthesized in the skin under the effect of ultraviolet radiation in the 296-310 nm wave length range.\footnote{1}

On the basis of aetiology rickets can be divided into two main groups.\footnote{2,3} The first one or “Classical” rickets is due to deficiency of Vit. D resulting from the lack of exposure to sunlight and inadequate dietary intake. The second or non deficiency types of rickets results from multiple aetiologies like chronic diarrhoea associated with fat malabsorption, hepatobiliary disease, chronic renal disease, disorders of renal tubular functions and use of anticonvulsants drugs like phenobarbete and phenytoin. Sometimes rickets is found as a part of rare conditions like proximal renal tubular acidosis and rare syndromes like de Toni Fanconi and Di George.

Classical rickets due to Vit D Deficiency (VDR) is a relatively common nutritional disorder in the paediatric population. As a nutritional disorder it is widely prevalent micro nutrient deficiency state among infants and children second only to iron deficiency anaemia in this country. However, lack of information to show the true incidence makes rickets a low profile disorder in Pakistan.

The high incidence of Rickets in Asians, in particular Pakistani immigrants living in UK has been mentioned in several studies Ford et al reported a 45% incidence of rickets in Infants in Bradford\footnote{4}, while Goel et al found that five to 7.5% of school going children in Glasgow had florid or subclinical rickets respectively.\footnote{5}

Regarded as an old disease, rickets has been overshadowed by the new and more exotic micronutrient deficiency disorders including Vit A and Zinc. This bias is clearly reflected in the fact that with the exception of few standard publications\footnote{6,7,8} there has been a shortage of published data available on rickets in this country. The country report on nutrition as well as national nutrition surveys (NNS) do not mention nutritional rickets as an important nutritional disorder.

Rickets received great attention as a clinically important nutritional disorder in Europe after the first world war as the after effect of industrialization and resultant rapid urbanization. This was attributed to the emergence of multistory buildings in the large industrial towns in the northern hemisphere where people lived under cloudy and foggy environment. However the improvement in the knowledge about the aetiology of rickets, and improved living standards backed by effective preventive strategies have resulted in virtual eradication of this disorder among the indigenous western populations. However it still remains an important clinical problem among the inmigrant populations living in these countries as well as among the inhabitants living in extremely Northern latitudes.\footnote{9,10}

The myriad of variable clinical presentation\footnote{11,12} makes rickets a less common diagnosis in the clinics of busy general physicians and paediatricians despite being a widely prevalent clinical disorder, causing significant morbidity among infants and young children, rickets is often missed clinically because of its non classical presentation. It is usually overlooked by the health planners as an insignificant public health problem. The notion that rickets is not an important nation-wide problem because there is abundant sunshine, may be responsible for treating rickets as a low profile disorder in Pakistan. However the facts and figures of this study show beyond any doubt that rickets is a widely prevalent disorder with high morbidity among the infants and children in this country, and as such it merits proper attention of both the clinicians and policy makers.
AIM OF STUDY

All infants and children (age group 6 months to 12 years approximate) attending the consultant outpatient paediatric clinic at Hayat Shaheed Teaching Hospital, Peshawar between January 1, 1995 till December 31, 1996 were clinically screened. Those having symptoms or signs suggestive of Rickets were assessed by detail history and complete clinical examination including anthropometric measurements.

After careful clinical evaluation the following were excluded from the study.

- Those who were born before 37 completed weeks of gestation (Pre term).
- Those who were graded having second or third degree malnutrition according to modified Gomez classification.
- Those who were mentally retarded.
- Those who suffered from serious systemic disease.

Those included in the study were registered and entered on a standard proforma after obtaining consent.

Information regarding the birth early feeding and weaning practices, living conditions socioeconomic background, type of housing, developmental and medical history including the use of drugs, was obtained and entered into the proforma. Radiographs of the wrist joints including the distal ends of Ulna and Radius were obtained. All those included were subject to haematological and biochemical investigations. These included complete blood picture total and differential white cells counts, serum calcium, serum alkaline phosphatase, inorganic phosphate, urea and electrolytes determination. The data was processed by computer analysis at the end of the study.

RESULTS

A total of 168 cases of Rickets were studied during the period of one year between January 1st 1995 and December 1996. Patients were divided into two main groups based on the basic cause (Fig. 1) and

<table>
<thead>
<tr>
<th>TABLE - I</th>
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<tbody>
<tr>
<td>SEX DISTRIBUTION AND MALE/ FEMALE RATIO IN 124 CASES OF RICKETS</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sex</th>
<th>Number of cases</th>
<th>Percentage</th>
<th>Mean age of presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>77</td>
<td>62%</td>
<td>14 months</td>
</tr>
<tr>
<td>Female</td>
<td>47</td>
<td>38%</td>
<td>23 months</td>
</tr>
</tbody>
</table>

![Fig. 1](image1)

![Fig. 2](image2)
TABLE – II
DEGREE OF SEVERITY OF RICKETS IN 124 CASES OF RICKETS

<table>
<thead>
<tr>
<th>Severity of rickets</th>
<th>Number of cases</th>
<th>Percent age</th>
<th>Ratio of mild florid rickets</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild and Moderate rickets</td>
<td>38</td>
<td>30%</td>
<td>2.3%</td>
</tr>
<tr>
<td>Floride rickets</td>
<td>86</td>
<td>70%</td>
<td></td>
</tr>
</tbody>
</table>

subgrouped according to the severity of the condition into clinically mild or moderate and florid rickets (table 2).

The majority 124(73.8%) cases were due to Vit D deficiency Rickets, while the remaining 44(26.2%) belonged to the non deficiency group.

The diagnosis of rickets was based on clinical and radiological criteria, supported by the evidence of abnormal biochemistry (table 3).

All the patients included in the study had some degree of radiological changes, pathognomonic of rickets such as metaphyseal splaying, fraying, cupping, widening of the epiphyseal growth plate and poor mineralization of the trabecular bone.

The most consistent biochemical abnormality was raised alkaline phosphatase, this was present in almost all the cases (table 4-A). Average serum alkaline phosphatase was 946 IU/L (upper normal limit -720 IU/L). The average serum calcium was 8.46 mg% (table 4-B) and inorganic phosphate was 3.44mg% (table 4-C). Blood urea and urine examination were normal in all of these children.

Among the Vit D deficient group 77(62%) were male and 47(38%) were female. Age distribution was different in male and female children. In males the mean age of presentation was 14 months (range 4-60 months). Female children with Rickets presented significantly later at an average age of 23 months (range 8-60 months) (table-1).

The clinical pattern of presentation was extremely variable in severity as well as quality. Forty eight % presented with recurrent episodes of chest infections 60% of these patients had wheeze in addition (Fig. 2).

Floppiness, with resultant delayed motor development was noted in 28% of the cases. 22% of the patients had diarrhoea with abdominal distention. Neurological manifestations like seizures, breath holding attacks and tetany were present in 10% and less than 10% of the cases respectively.

Seventy percent of the patients had one or more skeletal changes suggestive of rickets like metaphyseal sclerosis, cranio-tabes, ricketty rosary, pigeon chest deformity, genu varum or coxa vara present at the time of diagnosis.

Evaluation of the data about the possible etiology of rickets showed that in the Vit D deficiency group 108/124 infants (87%) were breast fed exclusively. Only 16 case received milk supplements. The mean age of weaning was 8.5 months. In 29 cases weaning was started later than 12 months.

Analysis of the data regarding habitat and housing showed (Fig. 3) that approximately 47% of the patients in the Vit D deficiency group belonged to the old city of Peshawar. They were residing in multistory buildings in areas of the old city suffering from heavy environmental pollution due to dust and smoke. The remaining patients in

TABLE – III
BIOCHEMICAL DATA OF 124 CASES OF RICKETS (MEAN VALUES)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum Calcium</td>
<td>8.46 mg/dl</td>
</tr>
<tr>
<td>Serum inorganic phosphate</td>
<td>3.44 mg/dl</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>944 I.U/l</td>
</tr>
<tr>
<td>Blood Urea</td>
<td>22.3 mg/dl</td>
</tr>
</tbody>
</table>
TABLE – IV
BIOCHEMICAL DATA OF 124 CASES OF RICKETS

A Serum alkaline phosphatase
(Adjusted upper normal limit = 702.1 U/l)

<table>
<thead>
<tr>
<th>International Units per litre</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>702–800</td>
<td>25</td>
<td>20%</td>
</tr>
<tr>
<td>800–1000</td>
<td>59</td>
<td>47%</td>
</tr>
<tr>
<td>1000–1500</td>
<td>26</td>
<td>20.2%</td>
</tr>
<tr>
<td>1500–3000</td>
<td>9</td>
<td>8.8%</td>
</tr>
<tr>
<td>3000–5000</td>
<td>5</td>
<td>4%</td>
</tr>
</tbody>
</table>

B Serum Calcium
(Normal range 9–11 mg/dl: 2.5 mg/dl = 1 mmol/l)

<table>
<thead>
<tr>
<th>mg/dl</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>7–8</td>
<td>28</td>
<td>23%</td>
</tr>
<tr>
<td>8–9</td>
<td>83</td>
<td>67%</td>
</tr>
<tr>
<td>9–10</td>
<td>13</td>
<td>10%</td>
</tr>
</tbody>
</table>

C Serum Inorganic Phosphate
(Normal range 3.4–5.5 mg/dl: 3.1 mg/dl = 1 mmol/l)

<table>
<thead>
<tr>
<th>mg/dl</th>
<th>Number of cases</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.5–3.0</td>
<td>17</td>
<td>15%</td>
</tr>
<tr>
<td>3.0–3.5</td>
<td>75</td>
<td>60%</td>
</tr>
<tr>
<td>3.5–4.5</td>
<td>27</td>
<td>11%</td>
</tr>
<tr>
<td>4.5–5.5</td>
<td>5</td>
<td>4%</td>
</tr>
</tbody>
</table>

DISCUSSION

This study was not designed to present the true prevalence of rickets among the population of infants and children in the NWFP. However, the study highlights rickets as a widely prevalent condition, evidenced from the fact that a fairly large number of cases were registered at a single paediatrics unit in Peshawar over the one year period of study. Inclusion of the other paediatric units in this study may have revealed the number of rickets patients to be truly substantial reaching almost epidemic proportions. Henceforth the present study is like the tip of the iceberg in comparison to the real magnitude of rickets in the community. This fact has been highlighted in a recent paper by researchers at Aga Khan Hospital.1

The preponderance of male infants and children in the study with early presentation of rickets (table 1) probably reflects the gender bias inherent to our culture.

Since in this study the majority (73.8%) of cases were found to be due to Vit D deficiency or classical rickets, therefore importance of Vit D deficiency in our infants and children population as a widely prevalent nutritional deficiency needs to be recognized and effective strategies for prevention evolved.6,8,9,11

The widely different clinical pattern of presentation of rickets in this study highlights the difficulty in the diagnosis of rickets in early stages. Most of those infants in children who presented with recurrent chest infections were commonly diagnosed as having asthma or pulmonary tuberculosis and treated as such. This not only delayed the proper treatment of the underlying problem but also exposed the patients to the risk of potentially harmful side effects of these treatments.

Diarrhoea was found to be present in 22% of the patients on initial presentation. Since diarrhoea is a common illness among infants and young children therefore the
significance of this finding cannot be established. Abdominal distention due to flabby musculature resulting from muscle weakness was noted in the majority of patients with or without diarrhoea.

Those who presented with generalized hypotonia due to metabolic myopathy causing delay in sitting, walking and other aspects of motor development were treated unnecessarily with brain tonics. Many of these unfortunate patients were labeled with the diagnosis of ‘cerebral palsy’ causing the parents serious mental stress.

Those presenting with seizures were diagnosed as epileptic or brain damaged. They were treated with anticonvulsants like phenobarbitone and phenytoine, with a potential to aggravate rather than treat the seizures occurring due to the metabolic disturbance associated with rickets.

The fact that approx. 47% (Fig. 3) of the cases recorded in this study belonged to the inner city slums of old Peshawar city, reflect therefore the significance of proper exposure to sunlight and the impact of environmental factors like smoke and dust to be considered in the overall situation analysis, before adopting preventive strategies against rickets. In the case of infants and children with rickets belonging to the rural regions of NWFP the negative impact of sociocultural factors and improper feeding / weaning practices is highlighted.

The cultural practices of cloddng the babies in heavy clothing in order to keep them warm denies the baby exposure to sunlight. The mothers of these babies who also spend most of their times indoors in purdah may be deficient in Vit D therefore the fastly growing baby does not get the required quantity of Vit D from these important sources. If introduction of weaning foods is delayed beyond age of 4-6 months and exclusive breast feeding continued then the development of rickets becomes a virtual certainty unless supple-

mented with Vit D. The fact that in this study majority of the babies were breast fed, confirms the observations of other researchers that exclusive breast feeding can not protect against rickets. It is possible that many of the lactating mothers are suffering from occult osteomalacia. The time of introduction of weaning food as well as the type of weaning diet also has important bearing on the occurrence of rickets. In this study many of the infants were put on a weaning diet after six months of age. Since most of our traditional weaning food consists of carbohydrates in the form of wheat based diets which contains high quantity of phytic acid, this may result in lowering the amount of available calcium and phosphorus for absorption. This fact has been mentioned in the studies conducted by Goel et al.

**CONCLUSION**

Vitamin D deficiency (including nutritional) Rickets is a widely prevalent but poorly recognised clinical disorder among the infants and young children in the NWFP. Diagnosis of rickets is often missed due to its extremely variable clinical presentation. Mild cases of Rickets are often not easily
diagnosed without a high degree of clinical suspicion pending biochemical confirmation. Therefore true magnitude of the problem is far greater than the apparent number of cases of florid rickets. Delay in diagnosis may lead to significant increase in acute morbidity as well as late complications seriously undermining the growth and developmental potential of the growing infant.

Practices like cladding the babies in dark clothes may deny the baby an important natural source of vitamin D. Therefore mere presence of sunlight is not protective unless some exposure is provided to both mother and baby. Breast feeding does not offer protection against rickets particularly if the mother is poorly exposed to sunlight due to sociocultural and climatic reasons. Delay in the introduction of semisolid food to the baby beyond four months of age and the selection of wheat based weaning foods are important contributing factors. Blanket Vitamin D supplementation to pregnant / lactating mothers and all the infants beyond four months of age up to 2 years and daily exposure to sunlight for at least two hours a day have proved to be important in the prophylaxis against nutritional rickets.

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