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ORIGINAL STUDY

The presentation of rickets to orthopaedic clinics : Return of the English Disease

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Rickets is a potentially treatable disease of the bone that is most commonly due to deficiency of vitamin D and is increasing in incidence in developed countries. Risk factors include dietary factors, the practice of covering up and darker skin pigmentation. This small retrospective case study set out to examine all cases of rickets presenting to the Paediatric Orthopaedic clinic over a 15-month period. Rickets presented in a bimodal fashion in the 6 cases identified : in males and females aged 3 or less and female adolescents aged 10 and above. This is in keeping with what is known regarding the rapid phases of growth during development. Five cases were from ethnic minority groups. Both female adolescents presented with genu valgum. Rickets can present primarily to Orthopaedic clinics with vague musculoskeletal symptoms. We recommend that biochemical screening be performed on patients from ethnic minorities who may be 'at risk'.

Keywords : rickets ; vit D deficiency ; risk factors ; biochemical screening.

INTRODUCTION

Rickets, a disease primarily caused by vitamin D deficiency, has been the subject of increasing focus in recent years. Originally described in the medical literature in the seventeenth century by the physician Whistler, the disease is believed to be re-emerging, particularly in the developed countries (*15*). The German for rickets is *'Englische Krankheit'* – the English disease, due to the prevalence of the disease in Victorian times.

In the U.K, several studies have indicated that vitamin D deficiency may be on the increase, particularly in Asian and African-Caribbean female populations in urban areas such as Manchester (*23*) and Birmingham (*11*). This is in keeping with the identified risk factors for vitamin D deficiency : darker skin pigmentations (*6*), the practice of covering up (*12*), prolonged periods of breast feeding by vitamin D deficient mothers (*9,25*) and a lack of usage of fortified "formula 'milk" (*29*) in more deprived minority groups.

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Blackburn with Darwen is an area in the county of Lancashire which has significant deprivation (*7*) and a sizeable ethnic population (*19*). The Royal Blackburn Hospital is the main hospital serving this area. The purpose of this study was to examine all cases of rickets presenting to the Paediatric Orthopaedic clinic over a 15-month period, including features and trends in its presentation. It is hoped that this will better inform the Orthopaedic Surgeon on the re-emergence of this disease.

PATIENTS AND METHODS

A retrospective case series study was performed on all patients diagnosed with rickets following presentation to the Paediatric Orthopaedic clinic at the Royal Blackburn Hospital between June 2007 and August 2008 (15 month period).

Demographic data regarding age, gender and ethnicity were collected, in addition to the presenting symptoms.

The diagnosis of rickets was made by using a combination of clinical, radiological and biochemical data. The latter comprised of testing for levels of parathyroid hormone (PTH), calcium and alkaline phosphatase (ALP). Vitamin D levels were also measured, unless it was felt that the diagnosis could be established on the basis of the other biochemical data. All patients were referred to the Paediatric department, which supervised the medical treatment of rickets.

RESULTS

Six cases of rickets in children were diagnosed following primary presentation to the orthopaedic clinic between June 2007 and August 2008. Five cases were from ethnic minority groups : 4 were South Asian and 1 Sudanese.

The demographics (age and ethnicity) of the patients, presenting symptoms and biochemical analyses are shown in Table I.

Case Histories

Case 1

An 11-month-old Asian male presented to the fracture clinic with a fracture of the mid shaft of his right radius and ulna secondary to being trodden on by his elder brother. Radiographs confirmed classical rickets : widening of the wrist growth plates and cupping of the metaphyses (Fig. 1). Biochemistry confirmed a low Vitamin D level and high Alkaline Phosphate and PTH levels. In addition he was iron deficient. The rickets was nutritional and responded to vitamin D supplementation. Fracture healing occurred successfully.

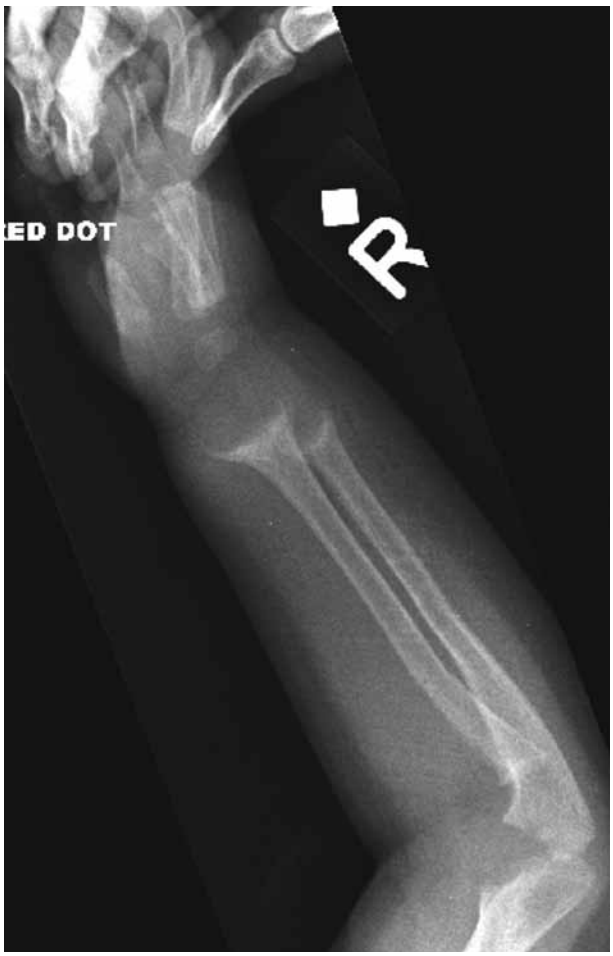

Fig. 1. — A radiograph of the right radius and ulna (case 1) demonstrating the characteristic widened growth plates and metaphyseal cupping of rickets.

Case 2

A 13-month-old Asian female was referred to the Orthopaedic clinic with swelling of the wrists and ribs. Radiographs confirmed the diagnosis of rickets. The PTH and Alkaline Phosphatase were

Table I. — Demographics, presenting symptoms and biochemical analyses of the patients diagnosed with rickets

Age	Ethnicity	Problem	Vit D 30-150 nmol/l	PTH 10-55 pg/ml	Ca 2.20-2.67 mmol/l	ALP 39-117 u/l
1. 11 mths	South Asian	Fracture forearm & swollen wrists	2.3	705	1.84	717
2. 13 mths	South Asian	Swollen wrists	11.9	1018	1.5	5818
3. 17 mths	Caucasian	External rotation of left foot & a valgus left tibia	13.6	17	2.32	165
4. 28 mths	Sudanese	Genu varum	22.8	1448	2.19	2582
5. 10 yrs	South Asian	Genu valgum	16.9	309	2.04	293
6. 14 yrs	South Asian	Genu valgum	–	400	2.06	390

massively raised (Table I). The serum calcium was lower than normal. Nutritional rickets was confirmed and treated by Vitamin D supplementation by the Paediatric department and the rickets resolved.

Case 3

A 17-month-old Caucasian male was referred by his General Practitioner to the paediatric Orthopaedic Clinic with asymmetrical out toeing. Clinically there was slight weakness of dorsiflexion of the left foot and a mild asymmetrical left sided valgus deformity to the tibia. His milestones were otherwise normal and he could walk. Radiographs were normal and MRI of the spine excluded a spinal cause. Bone biochemistry apart from a low Vitamin D level was normal. The Paediatricians confirmed nutritional Vitamin D deficiency and the neurological and musculoskeletal problems resolved with oral Vitamin D supplements.

Case 4

A 28-month-old Sudanese female presented with delayed walking and was not weight bearing. She was initially referred by the General Practitioner with limited hip abduction. Radiographs undertaken in the Paediatric Orthopaedic Clinic confirmed normal hip joints. There was evidence of previous stress fractures of the femur and widening of the wrist physes with cupping of the metaphyses. The biochemistry was highly abnormal (Table I.) The Paediatric Department confirmed and treated the nutritional rickets and the musculoskeletal abnormalities resolved.

Case 5

A 10-year-old female Asian presented with a progressive valgus deformity of the knees. The left valgus deformity (Fig. 2) was worse than the right with a mechanical index of 17° secondary to a hypoplastic lateral femoral condyle. Her bone biochemistry confirmed nutritional rickets which was treated by Vitamin D supplements (Table I). There were no associated neurological or myopathic abnormalities. The left knee valgus deformity was successfully corrected with a distal femoral opening wedge osteotomy (lateral approach, blade plate).

Case 6

A 14-year-old female Asian presented to the Paediatric Orthopaedic Clinic with a unilateral progressive valgus deformity of the left knee. There were no other significant symptoms. Radiographs confirmed a hypoplastic lateral femoral condyle with a mechanical valgus deformity of 16°. Biochemistry confirmed nutritional Vitamin D deficiency (Table I) which was treated appropriately. The femoral deformity was successfully treated by

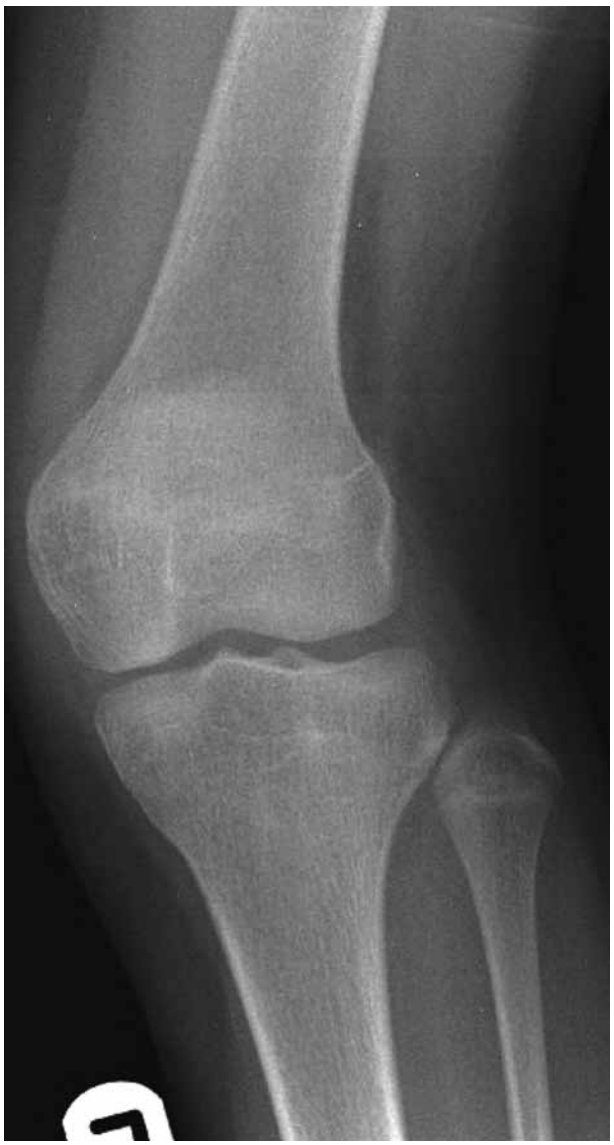

Fig. 2. — An antero-posterior (AP) radiograph of the left knee (case 5) illustrating the valgus deformity of rickets.

a distal femoral opening wedge osteotomy (lateral approach, blade plate).

DISCUSSION

There has been recent renewed interest in the medical literature (*21*) and the National press (*24*) in the re-emergence of Vitamin D deficient rickets in risk groups in the UK.

Vitamin D is responsible for the absorption of calcium in the gut and its use in the mineralisation of bone. It is primarily obtained either from dietary sources, or from synthesis in the skin through the action of sunlight, specifically ultraviolet B, on 7-dihydrocholesterol (*33*). Abnormalities in acquiring vitamin D from either of these two sources will therefore lead to its deficiency : human breast milk is low in vitamin D (*22*) and hence exclusive breast-feeding has been identified as a predisposing factor (*9*), particularly in mothers who themselves are vitamin D deficient (*25*). The consumption of phytate, a chelating agent to calcium found in high concentrations in chapatti flour, is of particular relevance in South Asian immigrant communities (*34*) and iron deficiency also has been implicated in the pathogenesis (*14*). Other dietary aetiologies include the practice of weaning off breast milk directly on to cow's milk, which has low vitamin D levels (*32*) and the low use of formulaic infant 'milks' that are fortified with vitamin D supplements (*32*). Few foods have useful levels of vitamin D naturally. Margarine is fortified (*4*) and oily fish have significant levels of Vitamin D (*5*). Many of the ethnic minority 'risk' groups may not eat these foods regularly (*4*).

The influence of sunlight exposure on vitamin D levels is well-established (*2,31*). Darker skin pigmentations affect the penetration of ultraviolet B and reduce the synthesis of vitamin D (*6*). The practice of covering up for cultural reasons (*12*) and the use of sunscreens (*16*) are also of relevance, particularly given recent trends in skin cancer awareness (*33*).

The clinical manifestations of vitamin D deficiency are varied and may include seizures, tetany (*10*), respiratory infections (*30*) and cardiovascular disease (*13*). In children, proximal myopathy (*8*) and cardiomyopathy (*28*) are recognised presentations. The skeletal features associated with rickets in children are believed to be due to an underlying failure of developing osteoid, or unmineralised bone tissue, to calcify (*33*). Long bone manifestations include genu varum in infants, or genu valgum in adolescents. Other abnormalities include swollen wrists, abnormalities of the costochondral joints and frontal bossing of the skull (*33*). Importantly

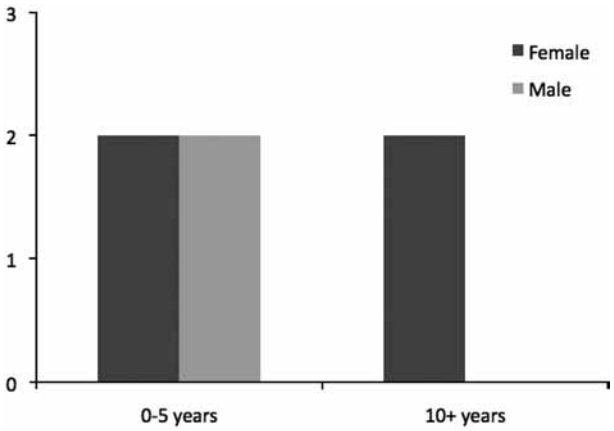

Fig. 3. — The distribution of females and males diagnosed with rickets presenting to the orthopaedic clinic from 2006-2008.

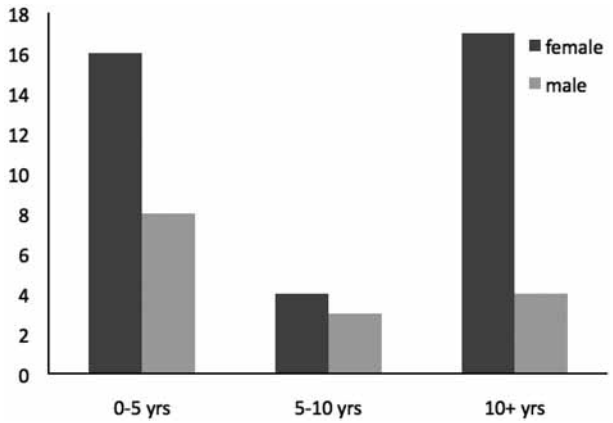

Fig. 4. — The distribution of females and males, by age, diagnosed with rickets, that presented to the department of paediatrics from 2003-2005 (*27*).

there is evidence to suggest that biochemical and radiological resolution of rickets is possible in younger children following treatment with vitamin D (*1*).

The data illustrates a bimodal distribution (Fig. 3) in the presentation of rickets and this is consistent with the findings in a larger series of 56 cases of rickets presenting to the Department of Paediatrics at the Royal Blackburn Hospital from 2003-2005 (*27*). This was a group of patients mainly of South Asian origin who presented both urgently and non-urgently with a wide variety of symptoms (Fig. 4). The skeletal abnormalities could be explained by the rapid phases of growth which have been shown to occur in the two age groups mentioned (*26*) as increasing metabolic demands for vitamin D during these times places an additional burden on pre-existing low vitamin D levels. This is a hypothesis shared by Moncrieff *et al* in a 1973 study (*18*).

An analysis of the demographics of Blackburn reveals a significant presence of several of the risk factors for vitamin D deficiency. According to the Indices of Deprivation 2007 (ID 2007), Blackburn with Darwen was ranked the 17th most deprived out of a total of 354 districts in England (*7*). The 2001 census revealed sizeable ethnic populations, for example, 10.66% were of Indian and 8.74% of Pakistani origin (*19*). In the same census, 21.23% of the population were Muslim (*20*), a religion in which the practice of covering up may be present. This was highlighted in the National press by the local MP for Blackburn, Mr Jack Straw (*3*). During 1971-2000, the county of Lancashire (containing Blackburn) received 50% and 32% of the average number of sunlight hours received by the Isle of Wight in the months of December and June, respectively (Met Office data (*17*)).

Biochemical screening for rickets may meet accepted screening criteria (World Health Organisation (*35*) (WHO)) and such a targeted programme may be worthwhile in those children falling within 'risk groups.' Such a decision is within the realms of the public health departments and should be combined with effective educational programmes in order to change certain 'risk' factors i.e. the fortification of certain foods with Vitamin D.

CONCLUSION

Vitamin D deficient rickets is a treatable metabolic disease, which is believed to be endemic in susceptible ethnic minority groups in the U.K due to clear risk factors. It is important that Orthopaedic Surgeons are aware of the re-emergence of this condition which has a bimodal distribution. Symptoms may be vague or dramatic. Severe deformity or proximal myopathies may be the presenting feature.

Long term serious health issues and skeletal deformities may develop, if rickets is not diagnosed and treated promptly.

REFERENCES

1. **Agarwal A, Gulati D.** Early adolescent nutritional rickets. *J Orthop Surg (Hong Kong)* 2009 ; 17 : 340-345.
2. **Brot C, Vestergaard P, Kolthoff N *et al*.** Vitamin D status and its adequacy in healthy Danish perimenopausal women : relationships to dietary intake, sun exposure and serum parathyroid hormone. *Br J Nutr* 2001 ; 86 Suppl 1 : S97-103.
3. **Browne A.** I would prefer women not to wear the veil at all, says Straw. *The Times*. 2006 07/10/2006. www.timesonline.co.uk/tol/news/politics/article664168.ece
4. **Calvo MS, Whiting SJ, Barton CN.** Vitamin D intake : a global perspective of current status. *J Nutr* 2005 ; 135 : 310-316.
5. **Chen TC, Chimeh F, Lu Z *et al*.** Factors that influence the cutaneous synthesis and dietary sources of vitamin D. *Arch Biochem Biophys* 2007 ; 460 : 213-217.
6. **Clemens TL, Adams JS, Henderson SL, Holick MF.** Increased skin pigment reduces the capacity of skin to synthesise vitamin D3. *Lancet* 1982 9 ;1(8263) : 74-76.
7. **Communities and Local Government.** The English indices of deprivation 2007 [database on the Internet]. 2008 [cited 07/10/2010]. Available from : www.communities.gov.uk/documents/communities/pdf/733520.pdf
8. **Crocombe S, Mughal MZ, Berry JL.** Symptomatic vitamin D deficiency among non-Caucasian adolescents living in the United Kingdom. *Arch Dis Child* 2004 ; 89 : 197-199.
9. **Dawodu A, Agarwal M, Hossain M, Kochiyil J, Zayed R.** Hypovitaminosis D and vitamin D deficiency in exclusively breast-feeding infants and their mothers in summer : a justification for vitamin D supplementation of breast-feeding infants. *J Pediatr* 2003 ; 142 : 169-73.
10. **Felner EI, Marks JF, Germak JA.** A variation of vitamin D deficiency in children. *J Pediatr Endocrinol Metab* 2001 ; 14 : 203-206.
11. **Ford L, Graham V, Wall A, Berg J.** Vitamin D concentrations in an UK inner-city multicultural outpatient population. *Ann Clin Biochem* 2006 ; 43 : 468-473.
12. **Gannage-Yared MH, Chemali R, Yaacoub N, Halaby G.** Hypovitaminosis D in a sunny country : relation to lifestyle and bone markers. *J Bone Miner Res* 2000 ; 15 : 1856-1862.
13. **Ginde AA, Scragg R, Schwartz RS, Camargo CA Jr.** Prospective study of serum 25-hydroxyvitamin D level, cardiovascular disease mortality, and all-cause mortality in older U.S. adults. *J Am Geriatr Soc* 2009 ; 57 : 1595-1603.
14. **Grindulis H, Scott PH, Belton NR, Wharton BA.** Combined deficiency of iron and vitamin D in Asian toddlers. *Arch Dis Child* 1986 ; 61 : 843-848.
15. **Holick MF.** Resurrection of vitamin D deficiency and rickets. *J Clin Invest* 2006 ; 116 : 2062-2072.
16. **Matsuoka LY, Ide L, Wortsman J, MacLaughlin JA, Holick MF.** Sunscreens suppress cutaneous vitamin D3 synthesis. *J Clin Endocrinol Metab* 1987 ; 64 : 1165-1168.
17. **Met Office.** Regional mapped climate averages 1971-2000 [database on the Internet]. [cited 03/10/2010]. Available from : http ://www.metoffice.gov.uk/climate/uk/averages/regmapavge.html
18. **Moncrieff MW, Lunt HR, Arthur LJ.** Nutritional rickets at puberty. *Arch Dis Child* 1973 ; 48 : 221-224.
19. **Office for National Statistics.** 2001 census : ethnic group [database on the Internet]. 2004 [cited 07/10/2010]. Available from : http://www.neighbourhood.statistics.gov.uk/dissemination/LeadTableView.do?a=3&b=276819&c=blackburn+with+darwen&d=13&e=16&g=388684&i=1001x1003x1004&m=0&r=1&s=1286481740843&enc=1&dsFamilyId=87
20. **Office for National Statistics.** 2001 census : religion [database on the Internet]. 2004 [cited 07/10/2010]. Available from : http://www.neighbourhood.statistics.gov.uk/dissemination/LeadTableView.do?a=3&b=276819&c=blackburn+with+darwen&d=13&e=16&g=388684&i=1001x1003x1004&m=0&r=1&s=1286481740859&enc=1&dsFamilyId=95
21. **Pearce SH, Cheetham TD.** Diagnosis and management of vitamin D deficiency. *BMJ* 2010 ; 340 : b5664.
22. **Reeve LE, Chesney RW, DeLuca HF.** Vitamin D of human milk : identification of biologically active forms. *Am J Clin Nutr* 1982 ; 36 : 122-126.
23. **Roy DK, Berry JL, Pye SR *et al*.** Vitamin D status and bone mass in UK South Asian women. *Bone* 2007 ; 40 : 200-204.
24. **Rose D.** TV and computer games blamed for return of rickets (newspaper article). *The Times*. 22/01/2010 [cited 07/10/2010]. Available from : www.timesonline.co.uk/tol/life_and_style/health/child_health/article6997656.ece
25. **Specker BL.** Do North American women need supplemental vitamin D during pregnancy or lactation ? *Am J Clin Nutr* 1994 ; 59(2 Suppl) : 484S-90S ; discussion 90S-91S.
26. **Tanner JM, Whitehouse RH.** Clinical longitudinal standards for height, weight, height velocity, weight velocity, and stages of puberty. *Arch Dis Child* 1976 ; 51 : 170-179.
27. **Tatnall S, Benjamin A, Qureishi S, Smith C.** Rickets in Blackburn : a public health concern. *Manchester Paediatric Club* ; 2006 28/09/2006 (presentation).
28. **Uysal S, Kalayci AG, Baysal K.** Cardiac functions in children with vitamin D deficiency rickets. *Pediatr Cardiol* 1999 ; 20 : 283-286.
29. **Wagner CL, Greer FR.** Prevention of rickets and vitamin D deficiency in infants, children, and adolescents. *Pediatrics* 2008 ; 122 : 1142-1152.
30. **Walker VP, Modlin RL.** The vitamin D connection to pediatric infections and immune function. *Pediatr Res* 2009 ; 65(5 Pt 2) : 106R-113R.

31. Webb AR, Kline L, Holick MF. Influence of season and latitude on the cutaneous synthesis of vitamin D3 : exposure to winter sunlight in Boston and Edmonton will not promote vitamin D3 synthesis in human skin. *J Clin Endocrinol Metab* 1988 ; 67 : 373-378.

32. Wharton BA. Weaning in Britain : practice, policy and problems. *Proc Nutr Soc* 1997 ; 56(1A) : 105-119.

33. Wharton B, Bishop N. Rickets. *Lancet* 2003 ; 362(9393) : 1389-1400.

34. Wills MR, Phillips JB, Day RC, Bateman EC. Phytic acid and nutritional rickets in immigrants. *Lancet* 1972 ; 1(7754) : 771-773.

35. Wilson J, Jungner G. Public health papers no. 34 : Principles and practice of screening for disease. World Health Organisation. Geneva : World Health Organisation ; 1968.