RICKETS

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FOREIGN LANGUAGES PUBLISHING HOUSE
Moscow

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РАХИТ

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PREFACE

Rickets was known as far back as early antiquity. Soranus of Ephesus (early 2nd century A.D.) described leg and spinal deformities in children. Galen (130-201 A.D.) described rachitic changes in the osseous system, including thoracic deformities, in greater detail.

In the 15th century this disease was quite widespread, especially in the large towns of North Europe. The Danish and Dutch painters of the time not infrequently portrayed children with thickened epiphyses, deformed chests, olympian foreheads, etc.

In the 17th century the children of England were widely affected with rickets. Descriptions of severe forms of this disease began to appear with increasing frequence in the works of English physicians and this fact warranted calling rickets the "English disease". The first work on the clinical aspects and anatomy of rickets, exhaustive for the time, belongs to the pen of Francis Glisson, English physician (1650). By the beginning of the 19th century the clinical picture of rickets was already very well known.

Russian authors have made a great contribution to the study of this disease. In his *Pediatrics*, published in St. Petersburg in 1847, S. F. Khotovitsky not only described in detail the skeletal changes, but also indicated such concomitant symptoms of rickets as hidrosis, muscular flaccidity, retarded motor development, changes in the gastrointestinal tract, etc. At the same time he noted

that an abundance of farinaceous and sweet foods, limited physical exercise, insufficient sunlight and fresh air, as well as debilitating diseases, were factors greatly conducive to development of rickets.

In his dissertation "On the Question of the Pathogenesis of the English Disease" N. S. Korsakov (1883) had established the importance of calcium in the pathology of this disease long before any chemical studies of the mineral metabolism in rachitic children were made. A. A. Kisel wrote his dissertation on the pathoanatomical changes in the bones of growing animals under the influence of minimal doses of phosphorus. V. P. Zhukovsky's dissertation (1894) was a result of his studies of the development of the "English disease" ("rachitism and rachitic deformities") among the children of St. Petersburg's workers.

I. A. Shabad's monograph *Calcium in the Pathology of Rickets* (1909) is still of considerable interest as the first study of mineral metabolism. M. S. Maslov in his dissertation "On the Physiological Importance of Phosphorus for the Growing Organism" (1913) revealed the deep metabolic processes in the child's organism.

A considerable contribution to the studies of the etiology, pathogenesis and clinical aspects of rickets has been made since the Great October Socialist Revolution by Russian pediatricians. Methods of prophylaxis and treatment have been elaborated in the works of Y. M. Lepsky, M. S. Maslov, A. F. Tur, Y. F. Dombrovskaya, P. S. Medovikov, S. O. Dulitsky, I. V. Tsimbler and the workers of the clinics under their supervision.

The works published since the last war deal with special clinical aspects, course and treatment of rickets in the unfavourable external environment produced by the war.

Rickets was formerly believed to be a disease of the moderate zone of Europe and America, the central cold belts and highly humid areas. But this view is incorrect. Rickets occurs both in countries located in the middle

latitudes and in the cold North and the hot South. Depending on the character of feeding, the child's general regimen and external environment (in the broad sense of the word), the difference may consist in the extent to which the disease is spread, as well as the forms and severity of its manifestations.

Rickets is quite widespread in many countries. At the end of last century examination of children of the East End of London, inhabited mainly by workers' families, revealed rickets in 80 per cent of the cases. In 1915 symptoms of rickets were observed in almost 80 per cent of the English primary school pupils. According to official figures of the British Ministry of Health, 87 per cent of kindergarten children suffered from rickets in 1928. In the city of Sheffield (England) 78.8 per cent of the children between 3 and 6 months of age were affected with this disease in 1944.

In 1930-33 rickets was found in 49.8 per cent of white children and 87.7 per cent of Negro children in the State of Tennessee (U.S.A.) In 1937 this disease was observed in 91-98 per cent of pre-school children in some large American cities.

In the 1940's rickets was observed in 74.2 per cent of the children between 2 and 3 years of age living in the industrial cities of Germany. In Belgium 80-90 per cent of the children were affected with rickets. A similar or somewhat lower rickets incidence was observed in other countries (A. N. Antonov, S. O. Dulitsky, Y. M. Lepsky).

The incidence of rickets among children in tsarist Russia was extraordinarily high. In large industrial cities this disease affected almost all children, 80 per cent of them suffering from rickets in Moscow (A. A. Kisel) and 95 per cent in Petersburg (V. P. Zhukovsky).

After the Great October Socialist Revolution the incidence of severe forms of rickets diminished, although the disease continued to be considerably widespread.

Of the total number of children under observation of

infant health centres in 1927, 57.6 per cent had rickets in Moscow, 69-82 per cent in Leningrad (according to individual infant health centres) and 64.9 per cent in Odessa (Y. M. Lepsky). Examination of children between 2 months and 2 years of age in Baku in 1939 revealed symptoms of rickets in 62.3 per cent of the cases (S. I. Gajiyeva). By 1949 the number of rachitic children considerably diminished and the grave forms leaving skeletal deformities disappeared.

In the Soviet Union the unfavourable living conditions engendered by the war raised the incidence of rickets not only among infants, but also among pre-school and school children. The number of children with second and third degree rickets more than doubled.

Since the end of the last war the steady rise in material and cultural standards of the population, the child protection measures and the increasingly more effective methods of preventing and treating various diseases since the end of the last war have led to a sharp reduction in child morbidity and mortality in the Soviet Union. The incidence of rickets among children has diminished and its severe forms have become a rare occurrence.

Nevertheless, clinical aspects, treatment and prevention of rickets are still urgent questions, since this disease continues to play a big part in child pathology.

It can no longer be doubted that rickets is a factor that diminishes the reactivity and immunobiological properties of the child's organism, increases its susceptibility to various diseases and, consequently, leads to greater morbidity and mortality. It should be noted that the resistance of the child's organism to various diseases is lowered not only by grave and clearly marked forms of rickets, but not infrequently also by effaced forms with barely perceptible clinical symptoms.

N. F. Filatov attached enormous importance to rickets as one of the causes of high child mortality. In his letter to S. P. Botkin in 1888 he wrote: "I am inclined to be-

lieve that children who die in the second year of life and later almost all have or had rickets and it is therefore no less important to eliminate the causes of rickets than it is to control diarrhea in children" (*International Clinic*, 1888, No. 5, p. 319).

Unfortunately, not all medical practitioners take into account this great pathological effect produced on the child's organism by rickets. G. N. Speransky is undoubtedly right in saying that "although pediatricians have efficient ways and means of preventing and treating severe forms of rickets at their disposal, they are not doing enough to prevent this disease. Some physicians fail to eliminate the harmful influence of this disease on the organism of the growing child" when they administer vitamins or expose children to irradiation indiscriminately or unsystematically, and overlook the importance of mild and effaced forms of rickets (*Medical News*, 1953, p. 1).

The Soviet health services are striving to eradicate rickets. Elimination of this disease is an indication of rational feeding, good regimen and proper care of children.

Numerous facts extending our knowledge of the etiology, pathogenesis, clinical aspects and methods of preventing and treating rickets have been accumulated in recent years. The last monograph dealing with these questions, *Rickets and Tetany of Rachitics*, was written by Y. M. Lepsky in 1941 and was republished in 1945. The author of this book has undertaken to generalise the available literary data and describe her long observations and studies. If this monograph in any way adds to the existing knowledge of the pathogenesis, clinical aspects, prevention and treatment of rickets, and helps the medical practitioners to control this disease, which is so very harmful to the organism of the child, the author will consider her aim in some measure achieved.

ETIOLOGY

It has long been known that unhygienic conditions are conducive to rickets. In children living in dark, crowded houses and spending very little time outdoors and in the sun rickets develops sooner and runs a severer course. In countries located closer to the equator, where children stay naked outdoors all day long, the rickets incidence is much lower than it is in countries with a moderate and cold climate. Rickets is also observed less frequently in children of some of the nomadic peoples. However, examinations of children of Buryat nomads in 1931 showed 85 per cent to have rickets despite the fact that they could stay out in the fresh air and sunlight all they wanted (Y. Y. Granat and Y. Zgorzhelskaya). The reason for it is that, according to a Buryat custom, the children were not taken outside their dark tents until the second year of life.

Rickets can also be observed in children living in southern countries if they are deprived of fresh air and sunlight. Even high living standards do not prevent the disease if a proper regimen is not observed. In cases of indoor life rickets affects children living even in highlands (M. Kh. Ugrelidze), where, as is well known, there is intense ultraviolet radiation.

In 1919 Huldschinsky discovered the effective action of the mercury vapour lamp (artificial mountain sunlight) in the treatment of rachitic children. Since then insufficient

irradiation of children with certain rays of the solar spectrum has been considered the main etiological factor of rickets.

Ultraviolet rays do not pass through ordinary window panes and are blocked by the air if it contains dust, soot and various vapours. Hence it is clear why rickets affects children living in lowlands with a humid climate and those living under any geographic conditions, but kept indoors during the first year of life.

The light theory also explains the seasonal character of rickets incidence.

Rickets strikes the most easily and runs the gravest course in late autumn, winter and early spring since children stay indoors for a long time, and even when outdoors they are so heavily dressed that very few ultraviolet rays penetrate to their bodies. In winter, when the sun does not rise high and the sky is long overcast with clouds, the ultraviolet rays are retained in the atmosphere and scarcely reach the earth. Fewer ultraviolet rays reach the earth's surface during early morning hours and in the evening when the sun hangs low.

The second important cause of rickets is irrational feeding of infants.

The advocates of the alimentary theory believed the development of rickets to be caused by a deficiency of calcium in the food because the bones affected with this process contain less calcium than do normal bones. The authors endeavoured to prove this experimentally by depriving experimental animals of the necessary amounts of calcium. Histological examinations have shown the bones of these animals to have defects which were not typical of rickets.

It was subsequently established that the calcium deficiency in the rachitic child's organism was not due to insufficient consumption of phosphorus and calcium with the food, but to a disturbance in the process of their assimilation by the child's organism.

It is generally recognised that breast-fed children are less frequently affected with rickets and never have severe forms of this disease. In cases of mixed and, especially, artificial feeding the incidence of rickets is much higher and the disease is severer.

The difference in the assimilation of calcium and phosphorus salts may be one of the causes of this phenomenon. A healthy breast-fed baby assimilates about 70 per cent of the calcium and 50 per cent of the phosphorus consumed with the food (Orgler); in cases of artificial feeding only 30 per cent of the calcium (I. A. Shabad) and 20-30 per cent of the phosphorus are assimilated. The calcium-phosphorus ratio (1 : 1.3-1.5) in human milk is the most favourable for the assimilation of these substances. The phosphorus and calcium of human milk are well assimilated by the child if the milk it consumes is raw. But if the child is fed drawn-off and boiled human milk, less of the phosphorus, calcium, protein fraction and fat is assimilated and more of these mineral substances and fat is excreted through the intestinal tract. This is largely due to inactivation of the enzymes, especially lipase, as a result of boiling the milk (A. B. Fridman). The vitamin C deficiency observed in children fed boiled human milk (M. N. Bessonova) in its turn sharply diminishes the enzymatic activity of the gastrointestinal tract and the acidity of the gastric juice. The alkaline reaction and deficiency of fatty acids in the upper portions of the small intestine inhibit the formation of calcium soaps, as a result of which the absorption of calcium, and, consequently, of phosphorus is disturbed.

Feeding the child boiled donor's milk is thus not equivalent to breast-feeding and does not prevent development of rickets.

But these are not the only dietary defects which lead to development of rickets. Assimilation of phosphorus and calcium requires a definite protein-fat ratio and the presence of alkaline and acid valences in the food. Prevalence

of oxalates or calcium salts over sodium may considerably affect the process of calcium absorption. Such foods as potatoes, cereals, beans and peas sharply impair assimilation of calcium. A definite ratio of calcium compounds and fat in the food (1 g of fat per 0.04-0.08 g of calcium) is necessary because a fat deficiency leads to insufficient formation of calcium salts of fatty acids (soaps). Even with adequate consumption of readily soluble calcium salts the process of their absorption in the small intestine is disturbed and leads to hypocalcemia if the amount of bile in the intestinal tract is diminished. Calcium is found in foodstuffs not only in the form of carbonates, phosphates, oxalates and fatty acid compounds, but also in an adsorbed state with proteins. It follows that assimilation of calcium also depends on protein metabolism. Normal phosphorus metabolism requires not only a definite ratio of the calcium and phosphorus compounds in the foodstuffs, but also of proteins, fats and carbohydrates, because a number of organic compounds of phosphorus with intermediate products of metabolism of these most important substances is formed in the process of assimilation. That is why a one-sided milk or carbohydrate diet of an infant, deficient in vitamins A, D, C and group B, causes a disturbance in the mineral metabolism and is conducive to the development of rickets.

Many investigators noted that children living in the Arctic Zone with no sunlight for a considerable part of the year were never or relatively rarely affected with rickets. They ascribed this circumstance to the special dietary habits of the people who consume a good deal of fish and seal flesh and fat.

In 1918 Mellanby demonstrated in an experiment with dogs that cod-liver oil acted as an anti-rachitic because it contained a special vitamin. For some time it was believed that the anti-rachitic activity of cod-liver oil depended on vitamin A already known at the time. McCollum passed a stream of oxygen through cod-liver oil,

which inactivated vitamin A, and discovered that the oil retained its anti-rachitic action. Subsequent research discovered in the nonsaponifiable part of cod-liver oil another vitamin with strong anti-rachitic action—vitamin D. It was thus definitely established that foodstuffs possess the property of preventing and curing rickets mainly in virtue of the amount of vitamin D they contain.

As a result of further research it was discovered that some foodstuffs possessing no prophylactic or curative action in rickets acquired it under the influence of ultraviolet rays. On irradiation the sterols contained in the foodstuffs (cholesterol, ergosterol) form substances some of which are members of the vitamin D group. This made it clear that irradiation of the child with short-wave ultraviolet rays formed vitamin D (D_3) in its skin, which is the essence of the anti-rachitic action of the rays of sunlight or a mercury vapour lamp. This gave rise to a firm conviction that rickets was caused by a vitamin D deficiency.

In addition to the afore-mentioned causes great importance in the etiology of rickets is also attached to other factors. According to some observations prematurely born babies are particularly susceptible to rickets. This is due not only to the increased vitamin D requirements of such children, as a result of their rapid growth and development, but also apparently to the fact that the specific injuries to the fetus, responsible for the premature birth, weaken the child's organism and in an unfavourable external environment facilitate the development of the rachitic process (M. S. Maslov).

No small part in the emergence of rickets is played by the diseases the child has survived. Gastrointestinal disorders lead to disturbance of all forms of metabolism, including that of minerals. In such cases acidosis quickly develops and there is a deficiency not only of vitamin D, but also of vitamins A, C and B, which in their turn facilitate development of rickets. A similar pathological effect is produced by respiratory diseases, all acute infectious

diseases, etc. It has been demonstrated, both experimentally and clinically, that disorders of the endocrine functions are conducive to emergence of rickets.

It was formerly held that rickets could be congenital. This was asserted in works published at the end of the 19th and the beginning of the 20th centuries (Kassowitz, Marfan, Czerny, Ploos van-Amstel, et al). The authors believed the small thickenings on the ribs at the junction of bone and cartilage or the softening of the flat bones of the skull to be manifestations of rickets in the newborn. The development of congenital rickets was associated with intrauterine diseases of the fetus (P. Giraud, Louchet) and entirely groundlessly with a hereditary predisposition.

N. F. Filatov, P. S. Medovikov, P. V. Kuskov, F. Y. Chistovich, A. Razumovsky, G. N. Speransky, A. F. Tur, M. S. Maslov and Wieland deny the existence of congenital rickets. The changes in the bones (soft skull bones, pliable sutures, negligible swelling at the bone-cartilage junction of the ribs), most frequently observed in prematurely born children, on closer scrutiny, prove to be nonrachitic but connected with incomplete physiological ossification and usually disappear by the second or third month of life.

Of interest in this respect is the work of Z. G. Korolyova (1955). By observing 712 children from the moment of their birth she discovered soft skull bones in 71 of them at the age of 2-3 weeks. In some of them one parietal bone was soft, in others—both bones, the softening affecting a narrow line along the sagittal suture or on the parietal bones parallel to the lambdoid suture. In some children the softening affected vast sections of both parietal bones. Marked changes in the parietal bones were usually observed in prematurely born children, or in those who were underweight at birth, and disappeared by the end of the fourth or fifth week. Roentgenoscopy of the shin and forearm bones revealed no deviations from the

normal structure. The phosphorus and calcium content in the blood serum was within the norm for healthy children of that age. Pathomorphological examinations of dead children, who had soft skull bones when alive, revealed no signs of rickets. These data once more disprove the existence of congenital rickets.

In conclusion it may be said that rickets is a disease conditioned mainly by social factors, for which reason it gained currency first among children of the indigent sections of the population in large industrial towns. Low material and cultural standards of the family, poor housing conditions, violation of the rules of hygiene in caring for the child, insufficient use of fresh air and sunshine and lack of regular physical exercise are all factors conducive to rickets. The character of feeding—one-sided, inadequate diet with insufficient vitamins A, D, C and B—is, as was already mentioned, particularly important. The health of the child's mother also plays a big part. The diseases suffered by the woman during pregnancy may essentially affect the child's development, weaken its organism and facilitate the subsequent emergence of rickets.