

## FURTHER OBSERVATIONS ON THE KENHARDT BONE DISEASE AND ITS RELATION TO FLUOROSIS

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In a previous report<sup>1</sup> my co-workers and I described 2 cases of a strange bone disease occurring in a Coloured location (Rooiblok) at Kenhardt in the north-eastern Karoo. We concluded that this condition — manifesting particularly as pain in the shins with later bowing of the femora and tibiae — was related to the high fluorine content of the local drinking water, and affected a large proportion of the schoolchildren from that location. White children and those Coloured children who lived in a different location in Kenhardt were not affected. The teeth of the 2 children we saw were severely fluorotic. X-rays of the pelvis of both parents of one child showed gross classical osteosclerotic fluorosis. We were unable to find any description in the literature of an affection of the bones similar to that found in these children, and were unable to determine what additional factor or factors had so modified the skeletal effects of fluorine that they occurred at a young age and started with rarefaction and softening instead of sclerosis.

In August 1961 we visited Kenhardt itself to obtain further information, which forms the body of this report. In the meantime the problem had been studied on the spot by a team from Pretoria University Medical School,<sup>2</sup> and I shall quote from its report where appropriate. In this report, mention is made of similar skeletal deformities occurring together with a high fluorine content in the drinking water in the Potgietersrus district. No further information is given.

### HISTORY AND TOPOGRAPHY

Before the 'great flood' of 1941 it is known that White families lived in the Rooiblok, including some 30 or more children. Apparently no leg pains or deformities were known to occur in these families (some members of which have been seen by us), despite the fact that their water was obtained from shallow wells only about 10 feet in depth. Their teeth, however, were reputed to have been brown and pitted, and to have crumbled away at an early age.

In 1939 Ockerse,<sup>3</sup> a dental health officer, investigated the condition of children's teeth in Kenhardt and nearby towns. Fluorosis of the teeth was more advanced in Kenhardt than elsewhere. Some of his findings are summarized in Table I.

Ockerse concluded that the differences in the prevalence of dental caries were caused by differences in the composition

TABLE I. FLUORINE AND CHILDREN'S TEETH (FROM OCKERSE<sup>3</sup>)

Town	No. of children examined	% with mottled enamel	% with caries	Amount of fluorine in water (mg. per litre*)	Amount of calcium in water (mg. per litre)
Upington	767	15	76	0.38	45
Kenhardt	318	82	26	6.8	67
Pofadder	183	94	17	2.5	153

\* 'Mg. per litre' and 'parts per million' (ppm) are identical.

of the drinking water. He stated that the severity of the fluorotic effect on the teeth in Kenhardt and Pofadder would itself predispose to caries. Nevertheless, the incidence of caries in the low-fluorine area of Upington was far greater (Table I). Admittedly the calcium content of the water was lower there, but it was little different from that at Kenhardt, and this was probably unimportant.

After the flood of 1941, Rooiblok was divided transversely into 21 long narrow plots of land and was settled by Coloured people. They dug wells in these plots about 25-30 feet deep and operated them by means of windmills. These wells were above the topmost rock-bed which occurs at about 60 feet.

Early in 1961 two floods occurred which largely devastated the Rooiblok, so that roughly two-thirds of the previous population of 581 had to move elsewhere, mostly to the other location. In this way, by an act of God, the problem of supplying them with less evil water was partly solved!

The main geological formation in the Kenhardt area, according to the geological survey of the Department of Mines, is old granite and gneisses. As previously described<sup>1</sup> there are 4 main dwelling areas at Kenhardt — the town (population 843 at the 1960 census), the Lokasie (Coloured population 1,253), Rooiblok (Coloured population 581), and along the course of the (usually dry) overflow river from the Rooiberg dam ('Rooibergskema', mostly Coloured, total population 149).

### Water Supply

In the town the fluorine content of the water in 1939 was noted as being high, namely 6.8 parts per million (ppm). The town water is supplied from comparatively shallow wells, sunk to a depth of about 50 feet. An alternative source of water, which can be used only in times of good rainfall, is obtained from shallow drifts at varying depths of 8-30 feet, which drain radially into a central collecting system. This water contains 1-2 ppm of fluorine.

The water of the Rooiberg dam, used so far only for irrigation purposes by certain people with 'rights', is comparatively low in fluorine content.

Some results of recent analyses follow (expressed as parts per million):

February 1960 (Mr. N. Penny, Government Pathology Laboratory, Cape Town):		Fluorine ppm	
Kenhardt town (tap)	....	2.6	
Rooiblok North	....	6.0	
Rooiblok South	....	7.4	
Rooiblok South (September's well)	....	11.6	
February 1960 (University of Pretoria):		Fluorine ppm	
Kenhardt town (various)	....	2.6-3.2	
Rooiberg dam (dry bed)	....	1.8	
Rooibergskema	....	4.0	
Rooiblok (North, various)	....	3.6-8.0	
Rooiblok (South, various)	....	7.5-13.0 (highest content from September's well)	
March 1960—before the recent floods (Skeikundige Laboratorium):		Fluorine ppm	Calcium carbonate ppm
Kenhardt town (from borehole)	....	4.2	300
Rooiblok South (September's well—most evil)	....	4.8	420
Rooiblok Central	....	3.7	330

Several complete water analyses are available, but do not appear to be worth recording, since they show no outstanding abnormalities. In case some rare element was playing a part, a semi-quantitative analysis of 4 specimens of water was made by mass spectrograph in 1960 (courtesy of Western Province Fruit Research Station). The water from South Rooiblok (September's well) and Central Rooiblok differed slightly from that of the town and from a specimen from North Rooiblok in being rather richer in boron, lead and aluminium. Ordinary analysis of the same water for calcium gave between 75 and 90 mg. per litre (identical to ppm) in Rooiblok and only

38 ppm in the town water. Full ordinary analyses were as follows:

	Town water	N. Rooiblok	Central Rooiblok	S. Rooiblok (most evil)
pH	7.9	7.3	7.5	6.9
Na (ppm)	168	421	375	340
K	2.3	7.1	2.2	5.5
Ca	76	79	38	90
Mg	33	66	26	49
Cl	200	475	360	450
SO <sub>4</sub>	122	335	133	115
HCO <sub>3</sub>	324	384	418	427

Other analyses for fluorine include:  
 Bone biopsy from Stoffel September (1960): 8,500 ppm (calculated on bone ash)  
 Sheep bone from Rooiblok (1960): 145 ppm  
 Goat horn from Rooiblok (1961): 2,200 ppm

SURVEY OF KENHARDT INHABITANTS

Methods

The Coloured and European schools were visited (by courtesy of the headmasters, Mr. von Freda and Mr. Perold.) In the Coloured school there were 540 pupils between the ages of 6 and 14, of whom some 200 came from Rooiblok. All pupils in standard I (aged 7-10), and a similar group in the European school, were examined. Some additional children were examined in the Rooiblok location itself. The appearance of the teeth was graded as follows:

- O = no evidence of fluorosis;
- ± = doubtful;
- + = definite, occasional, opaque white or brown flecks;
- ++ = many white or brown flecks or streaks with early pitting;
- +++ = severe brown discoloration with much pitting, often caries at gingival margin; and
- ++++ = brown teeth, reduced in size by actual crumbling away of the free margins.

The height, weight and age of each child, and their dwelling history, with exact source of drinking water, were ascertained. The standard of food intake of the Coloured children was roughly assessed with the help of the local schoolteachers. Evidence for, or age of, puberty was noted, together with any complaints of pain in the back or legs, or any deformity of the legs or defective movement. Entries were made on special proformas.

Selected X-rays, including in all schoolchildren an A-P view of one femur and lateral view of one tibia, were taken later, and assessed by myself in collaboration with Dr. L. Werbeloff (Senior Radiologist at Groote Schuur Hospital). In addition, several complete school classes were lined up, questioned as to whether they suffered pains in the legs on walking or playing games, or at other times, and briefly inspected for obvious bony deformity and goitre. A number of adults, both Coloured and European, were also examined.

Teeth

It can be noted from Tables II and III that the teeth of White children and of Lokasie Coloured children were very similarly affected. Both groups drank the same water. Plainly Rooiblok children showed more advanced dental fluorosis. Severely affected teeth were seen in children who drank at quite different wells in the Rooiblok location. The teeth of 7 children who used September's

TABLE II. DEGREE OF TOOTH FLUOROSIS IN WHITE SCHOOLCHILDREN

	0	±	+	++	+++	++++
Always lived in Kenhardt (25*)	2	3	14	6	0	0
From outlying areas (25)	10	6	8	1	0	0
Total (50)	12	9	22	7	0	0

\* Figures in parenthesis in Tables II, III, IV, V and VII indicate the numbers of children in each particular group.

TABLE III. DEGREE OF TOOTH FLUOROSIS IN COLOURED SCHOOLCHILDREN

	0	±	+	++	+++	++++
From Lokasie (town water) (32)	3	5	16	8	0	0
From Rooiblok (37)	0	1	12	11	10	3
Total (69)	3	6	28	19	10	3

well in South Rooiblok (the well highest in fluorine content) were affected thus: 3 one-plus, 1 two-plus, 2 three-plus and 1 four-plus.

Children of preschool age in the Rooiblok frequently showed marked fluorosis of their deciduous teeth—even under the age of 2. The youngest child we saw who had at least 2-plus fluorosis was aged 10 months.



Fig. 1. Schoolchildren from the Rooiblok with deformities of the lower limbs (original photographs by courtesy of Prof. D. Steyn).

### Bones

It was soon evident that we had already seen the most severe cases of bone deformity of the type previously described — lateral bowing of femora and anterior bowing of tibiae, with variable knock-knee or bow-leg. Two other severely affected children had already undergone osteotomy, and two reputedly similar persons had left the district (including the oldest known affected member, aged 25). Photographs of some of the affected children are shown (Fig. 1). Assessment of mild degrees of bony affection by physical examination proved quite unreliable, so that only radiographic evidence will be used here.

In addition to children with the bony deformities we were looking for we encountered a boy with a vitamin-D-resistant rickets, a girl with Blount's disease and a boy with a severe congenital unilateral valgus deformity at the knees.

### Bone Pain

No children complained of pain in the back.

White schoolchildren — 3 of 60 complained of pain in the legs (all in knees).

Coloured children from the Lokasie — 3 of 124 complained of pain in the legs (1 in knees, 2 in calves).

Coloured children from Rooiblok — 47 of 98 complained of pain in the legs (all in the shins).

### Radiographic Survey

The abnormalities seen in the X-rays of the leg bones (Fig. 2) included thinning of the cortex with widening of



Fig. 2. Three affected tibiae from Rooiblok compared with a normal bone (extreme right). Note general expansion of shafts, thinning of cortices and slight forward bowing.

the medullary cavity in the lower part of the femur and upper end of the tibia, forward bowing of the tibia with anterior thickening (buttressing) of the cortex in that area, and obliteration of the normally clear demarcation between cortex and medulla. Lateral view of the spine showed transverse (Harris) lines, and the bones of the hands of affected children were clearly demineralized, with coarse trabeculation.

It can be seen from Table IV that Lokasie children are probably completely unaffected, while the most severely affected are those from September's well in Rooiblok South. Other affected children in Rooiblok drank at various wells, including some who used wells with

TABLE IV. X-RAYS OF LEGS OF COLOURED SCHOOLCHILDREN

	Abnormal	Doubtful	Normal
Rooiblok children (31) .. .. .	15	4	12
Lokasie children (34) .. .. .	0	5	29
September-well children (9) * .. .	7	1	1

\* This group is also included in the Rooiblok children.

apparently rather low fluorine content (e.g. Julies' well with 3.6 ppm in North Rooiblok). No affected children (or children complaining of bone pain) were found coming from the banks of the 'overflow river' (Rooibergskema).

### Correlation between Tooth and Bone Abnormalities

As shown in Table V, there was little correlation between bony involvement and degree of tooth fluorosis, except

TABLE V. CORRELATION BETWEEN TEETH AND RADIOLOGICALLY ABNORMAL BONES IN ROOIBLOK CHILDREN

	Teeth				
	0	+	++	+++	++++
Bones normal (15) .. .. .	1	6	5	3	0
Bones abnormal (18) .. .. .	0	6	4	5	3

that all 3 children with crumbling teeth (4-plus) had bone disease. Table VI shows that there appeared to be no correlation between tooth affection and bone pain, but

TABLE VI. CORRELATION BETWEEN SHIN PAIN, BONY ABNORMALITY, AND TEETH IN ROOIBLOK CHILDREN

	Bone pain			No bone pain		
	Teeth	Teeth	Total	Teeth	Teeth	Total
	0 to	+++ to		0 to	+++ to	
Bones normal on X-ray .. .. .	6	0	6	7	3	10
Bones abnormal on X-ray .. .. .	6	4	10	4	3	7

a possible partial correlation between bone (shin) pain and radiologically abnormal bone (not quite statistically significant at 95% level).

### State of General Health

Many of the Coloured children did not give the appearance of being well-fed, but no specific deficiency diseases were seen. Steyn *et al.*<sup>2</sup> also remarked on the complete absence of clinical evidence suggesting deficiency of vitamin C. The typical diet included meat and milk once or twice a week, an occasional egg, and large quantities of 85% extraction bread, with hardly any green vegetables or fruit except oranges when in season.

A chronic nutritional deficiency is presumably responsible for the findings shown in Table VII, in which it is

TABLE VII. MEAN HEIGHTS IN DIFFERENT GROUPS (In inches)

	8 years old	9 years old
White schoolchildren .. .. .	52.9 (22)	55.5 (21)
Lokasie schoolchildren .. .. .	48.8 (20)	49.5 (15)
Rooiblok schoolchildren .. .. .	48.7 (4)	50.2 (7)

plain that the Coloured children were well below the Whites in mean height. There was, however, no discernible difference between the Rooiblok and other Coloured children.

No relationship was found between the general nutritional state and the assessment of food intake on the one hand and bone or tooth disease on the other.

Not one single goitre was seen in over 300 schoolchildren (though few necks were actually palpated). The local medical practitioners believe goitre to be very uncommon in this area.

Serological studies on 20 Rooiblok children, several with bone deformities, were all negative for syphilis (performed by Dr. T. Sacks of Pretoria University), as were the previous tests on the patients seen in Groote Schuur Hospital.

In view of the abnormal radiographic appearances, somewhat resembling those associated with a chronic haemolytic process (e.g. as in Cooley's anaemia), haemoglobin estimations were performed on 13 children from Rooiblok (several with affected bones). The results ranged from 12.5 to 16.5 G. per 100 ml. There was thus no anaemia.

#### Particular Cases

Six of the children with radiologically abnormal bones had been born outside Rooiblok and had moved there some 5 or 6 years previously. Bone pain apparently started after 3-4 years' residence in Rooiblok.

Several elderly Coloured people were questioned for back pain and examined for ability to flex the spine and touch their toes. No complaints of back pain were found, and flexion was remarkably good, even in those showing severe osteosclerotic fluorosis of classical type on X-ray. Thus the two old Septembers, who claim to have drunk from the same evil well since they were 21, could touch their toes with ease (Fig. 3), despite grossly fluorotic bones (Figs. 15 and 16 of



Fig. 3. The two old Septembers touching toes with straight knees despite severe sclerotic fluorosis (illustrated in Figs. 15 and 16 of our first paper).

previous communication<sup>1</sup>). The old man had some teeth left, which showed only one-plus fluorosis. The teeth of the goats belonging to the Septembers appeared normal.

A Coloured man aged 50, who had always lived at the Lokasie, had previously been diagnosed elsewhere as suffering from marble-bone disease with bilateral clavicular fractures. He actually had moderate fluorosis, with calcification of the coracoclavicular ligaments.

#### DISCUSSION

##### Possible Relation to Rickets

The occurrence of softened bones in childhood with lower-limb deformities compatible with old rickets, raises the possibility of this disease being present, at least in conjunction with fluorosis. All additional factors are against this, however:

1. All Coloured children in the Kenhardt area get plenty of sunshine.

2. Their diet is not low in phosphate; their calcium intake is certainly lower than approved standards, but it is no lower than is general in South Africa, and in any event there is no evidence that this factor ever leads to rickets.

3. Children coming into the Rooiblok, long past the ordinary rachitic age group, are liable to the same disorder. On the other hand, similar Coloured children, living in the other location one mile away, are unaffected.

4. The age of onset of the bony disorder is too late for ordinary vitamin-D-lack rickets.

5. Clinical rickets in infancy is unknown in the area.

6. No clinical or radiological evidence of active rickets was observed by us or by the Pretoria team.<sup>2</sup>

7. The 2 patients we examined at Groote Schuur Hospital and several others whose blood was taken by the Pretoria team showed no biochemical abnormalities.<sup>2</sup>

The single biopsy specimen we obtained did not show evidence of active rickets, but the inclusion of islands of cartilage in the bony substance (seen in biopsy) can be a feature of healed rickets. However, the balance of evidence strongly favours the thesis that the rachitic-like bowing and biopsy changes are non-specific and caused by agencies other than rickets.

##### Relation to Fluorosis

We have confirmed that the bone disorder of the children of Rooiblok is neither congenital nor inherited. It is neither rachitic nor syphilitic in nature, nor is it related to anaemia. It affects children from all parts of Rooiblok, but especially those who come from the south end, and most especially those who use September's well (plot No. 1284, penultimate plot from the southern end). It appears to require 3-5 years' exposure to evil waters for its genesis. It appears to have some relationship to the high fluorine content of the drinking water, because:

1. All affected people have drunk water with a high fluorine content.

2. The most affected, by and large, are those who have drunk from wells with the highest fluorine content.

3. Bone changes in a patient (Stoffel September) previously described in detail,<sup>1</sup> were intermediate between the 'osteoporotic' type and the classical sclerotic type. His bone biopsy was similar in some ways to that described in adult fluorosis<sup>4</sup> and it contained a large amount of fluorine. His parents had typical gross sclerotic fluorosis.

4. No case of abnormal bone changes was seen without at least mottling of the teeth.

However, fluorine cannot be the sole factor because:

1. Equally high fluorine intake in other parts of South Africa and other countries has not been reported as producing such a condition; in fact statements are made to the effect that 20-30 years' exposure to water with a high fluorine content is necessary for bone disease to occur, and then it is of the sclerotic type.<sup>5-7\*</sup>

2. There was little correlation between bony abnormality and the degree of fluorosis of the teeth.

\* In experimental animals, the early skeletal changes produced by fluorine may be 'osteoporotic' or 'osteomalacic', as discussed in detail by de Senarclens.<sup>8</sup> Apart from the Kenhardt phenomena, these findings appear to have no human counterpart.

3. Bone disease was found in relation to water with a comparatively low fluorine content (although this content might have varied from time to time).

#### *Relation to other Environmental Factors*

Further than this we are still unable to go. From the evidence so far adduced we appeared to be dealing with a variety of fluorosis which had been modified by some additional environmental factor. There was no evidence to suggest that this factor was a dietary one, although it could be argued that poor nutrition might enhance the deleterious effect of fluorine. The most probable source of the additional factor appeared to be the water supply, but we have been unable to discover any particularly outstanding abnormality in it, beyond the high content of fluorine.

On mass spectrographic analysis, two of the Rooiblok samples contained rather more lead, boron and aluminium than the town water, but the significance of this is not clear. (We found no clinical indication of chronic lead poisoning.) A third 'evil' water did not show this peculiarity.

The highest prevalence of bone and tooth disease and the most severe cases were related to one particular well, September's well, which also showed the highest fluorine content whenever tested. However, one or two children who had always lived on the Septembers' plot, completely escaped the bone disorder. This suggests differences in 'individual susceptibility'.

The calcium content of the water in the Rooiblok was not particularly low — making it most unlikely that these people were subjected to a much lower intake of calcium than their neighbours. The well-water used in the Rooiblok had considerably greater salinity than the town water; in fact this high salinity brought it to the borderline of potability. However it is difficult to understand how this might alter bodily reaction to fluoride.

#### *Relation to Pain and Disability*

The significance of the pain complained of in the shin is difficult to estimate, but the immediate response of 50% of the Rooiblok children to the question was very impressive, especially when compared with the lack of response of other sections of the community. It would certainly appear that the earliest feature of the bone disorder of Kenhardt is shin pain, and that this is very common indeed.

On the other hand it was surprising to observe the absence of pain or of impediment in movements of the spine and large joints in cases of radiologically severe osteosclerotic fluorosis. This condition, almost certainly quite common in all groups of the older inhabitants of Kenhardt, did not appear to produce any disability whatever.

#### *Future Water Supplies*

It appears that the present drinking water obtained from the Rooiblok wells is evil beyond its mere fluorine content. Many of the previous inhabitants have already moved, and now obtain their water from the town supply. The latter, however, has for years had a rather high fluorine content, except for occasional periods after good rains when it is derived from a different source, as mentioned above. Eventually it is planned to enlarge the Rooiberg

dam and so create a good standard water-supply for the whole town. In the meantime, at least the children remaining in Rooiblok should be supplied with water from elsewhere.

From the purely local evidence obtained from Kenhardt and outlined above, the presence of quite a high fluorine content (at least up to 3 ppm) produces no disability on its own and in fact appears to protect the teeth from caries, although in some people it may lead to a degree of unsightly brown staining. It would seem rather unnecessary to insist upon a fluorine content in drinking water of less than 3 ppm in those country areas where such water would be extremely difficult, if not virtually impossible, to obtain.

#### CONCLUSION AND SUMMARY

1. The bony disorder in children of the Rooiblok location of Kenhardt is related to the high fluorine content of their drinking water, together with some other environmental factor which we have been unable to determine.

2. The particular 'fluorosis' from which they suffer includes early and often severe affection of teeth, together with shin pain which may proceed to actual softening of the leg bones, with bowing.

3. Radiological changes showing demineralization, quite unlike those of classical sclerotic fluorosis, are common in the Rooiblok children, and may occur in the absence of bone pain or any other clinical feature.

4. Older people in the Kenhardt district may have radiographic evidence of classical osteosclerotic fluorosis, but without disability.

5. The high fluorine content of the water of Rooiblok and of Kenhardt itself does not appear to predispose towards gout, which is in fact rare.

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