Epidemiological evidence from Zaire for a dietary etiology of konzo, an upper motor neuron disease

T. Tylleskär, M. Banea, N. Bikangi, L. Fresco, L.Å. Persson, & H. Rosling

A clear association between seasonal outbreaks of a paralytic disease called konzo and toxic effects from consumption of insufficiently processed bitter cassava roots has been demonstrated in Bandundu region, Zaire. A community-based survey of 6764 inhabitants identified 110 live and 24 dead konzo-affected persons with a history of isolated non-progressive spastic paraparesis of abrupt onset. The start of these annual outbreaks of konzo in 1974 coincided with the completion of a new tarmac road to the capital, which facilitated the transport of cassava and made it the main cash crop. The extensive cassava sales encouraged the consumption by the peasant families of roots that had not been adequately processed; frequent acute cyanide intoxications resulted when the naturally occurring cyanogens in the roots were eaten. The disease mainly appeared in the dry season when there was high consumption of insufficiently processed cassava and the diet lacked supplementary foods with sulfur-containing amino acids which promote cyanide detoxification.

These results, which confirm the earlier findings in East Africa, show that, owing to the high cyanide and low sulfur dietary intake, there is an increased risk of konzo outbreaks in cassava-growing areas during periods of adverse agro-economic changes.

Introduction

Epidemic outbreaks and sporadic cases of spastic paraparesis have been reported from both the southern and central parts of Bandundu region in Zaire (1–5). The first report (I), in 1938, described an epidemic following a drought in the southern part of the region, where the disease was called “konzo”, a word in the local language describing the paralysis. A detailed neurological study in Bandundu (5) showed that this spastic paraparesis is identical with that reported from Mozambique (6). A recent report of the same type of spastic paraparesis in the United Republic of Tanzania (7) concluded that it was a distinct disease entity and should be named konzo. This disease is characterized by an abrupt onset of varying degrees of symmetrical spastic paraparesis due to isolated bilateral upper motor neuron damage. The disease always affects the distal upper motor neurons more than the proximal ones. Except for vision abnormalities and dysarthria in severe cases, only the pyramidal tracts are affected. Slightly affected persons may recover; in others the symptoms are permanent but non-progressive (7). Patients with konzo lack HTLV-I antibodies (7, 8); they also differ clinically from cases of HTLV-I-associated tropical spastic paraparesis, which is characterized by a gradual onset of a progressive spastic paraparesis (9, 10).

The acute onset of isolated pyramidal signs in konzo differentiates it clinically from most forms of tropical myeloneuropathies (11). The exception is lathyrosis (12), a spastic paraparesis of acute onset which is caused by high consumption of Lathyrus sativus, a legume that is not eaten by any of the populations affected by konzo.

Outbreaks of konzo in the drought-affected areas of East Africa have been attributed to several weeks of almost exclusive consumption of insufficiently processed bitter cassava roots (Manihot esculenta) (7, 13). Because of the high yield from poor soils, cassava has become the major staple crop in many parts of Africa (14). Roots from the high-yielding varieties of bitter* cassava may contain very

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*The terms “bitter” and “sweet” are in common use to distinguish, respectively, between cassava roots that need to be processed before consumption and those that can be eaten fresh or after simple boiling. The bitter roots contain high amounts of cyanogenic glucosides which are potentially toxic and give the bitter taste. Adverse growth conditions may increase the amount of glucosides in the roots.
high amounts of cyanogenic glucosides. These substances can be removed by efficient processing methods like soaking and other forms of fermentation (15). Too short a processing time may result in a high dietary cyanide. The toxic effects are aggravated by a low protein diet, especially a low supply of sulfur-containing amino acids which provide sulfur for the detoxification of cyanide to thiocyanate. A high cyanide and low sulfur dietary intake due to exclusive consumption of insufficiently processed cassava roots has been proposed as the main cause of konzo (7, 16).

Cassava is the main staple food of the konzo-affected population in Bandundu region, but an earlier study (5) failed to find any association between konzo and cassava toxicity. Based on the finding of clustering in families, an infectious etiology was suggested (5). The present study describes the epidemiology of konzo in a high prevalence area of Bandundu and examines the relationship with possible high consumption of inadequately processed bitter cassava.

Materials and methods

Study area

The Bandundu administrative region in Zaire (Fig. 1) lies to the east of the capital Kinshasa and covers 295,000 km² with a population of 3.7 million in 1984. The three administrative subregions have different agro-ecological characteristics. The northern one, Mai-Ndombe, north of the Kasai river, belongs to the Congo Basin rain forest and has a population density of about 5/km². The central subregion, Kwili, consists of grass-covered rolling hills with poor sandy soils intersected by widely forested, relatively more fertile river valleys running in a roughly south–north direction; the average population density is 25/km². The southern subregion, Kwango, constitutes a savanna tableland with some forested river valleys and has a population density of 10/km².

The annual rainfall in the region ranges from 2000 mm in the north to 1500 mm in the south. The main dry season (June to August) ranges from 70 days in the north to 115 days in the south. The population lives mainly in villages, growing cassava as the major subsistence and cash crop (17). The central part of the region has a relatively well-developed road network and some urban centres. More than 20 local languages are spoken in this ethnically heterogeneous region; Lingala is the lingua franca in Mai-Ndombe and Kikongo in south of the Kasai river.

Owing to the reported high prevalence of konzo (4) we selected the Lumbi Health Center catchment area (20 x 12 km), situated 30 km south of the administrative centre of Masi-Manimba in the central part of Bandundu, as the study area (Fig. 1).

Methods

The study was carried out in August 1988. With the informed consent and assistance of village leaders, a census was taken in all 21 villages in the area. The inhabitants were registered according to ethnic affiliation, sex, and age group (children were <15 years; adults, ≥15 years). The population in each village was screened for konzo by examination of all persons with locomotor disabilities who had been identified by the village leaders. The examinations were done in the villages by a physician (TT, BM or HR): gait while walking and running, regular use of one or two walking sticks, and speech abnormalities were noted. The size of the thyroid gland, presence of spinal abnormality, oedema or ulcers on the lower limbs, ankle clonus, and reflexes (plantar responses, knee and ankle jerks) were examined while in the

Fig. 1. The study area in Bandundu region in Zaire with the number of live and dead konzo patients as well as the number of inhabitants in each of the 21 villages, e.g., 35 alive + 13 dead and 1060 total = 35 + 13/1060.
sitting position. Vision and sensory abnormalities were investigated by questioning.

The criteria for konzo were a visible abnormality of gait while walking or running (symmetrically spastic), a history of abrupt onset (less than 1 week), a non-progressive course in a formerly healthy person, and bilaterally exaggerated knee and/or ankle jerks but without signs of disease of the spine. Persons fulfilling these criteria were interviewed in the local language according to a standardized questionnaire regarding the type of diet and time of onset. The year of onset was cross-checked with the dates on the birth certificates of children in the neighbourhood; the month of onset was determined by use of a local event calendar. Konzo-affected persons who had died were characterized through interviews with village leaders, neighbours, and relatives.

At Masi-Manimba hospital and Lumbi Health Center the clinical records were reviewed and the staff were interviewed regarding annual occurrences of konzo in the study area.

Focus group interviews (18) with about six adult participants of mixed ages were held in each village. The participants were selected by the local health staff at an announced information meeting in each village. Village leaders and others in positions of power were not included in the groups. The two to three hour discussions were held in private, gently guided by a well-trained nutritionist and a translator. Only one group interview was performed for the five westernmost villages, all situated closely together. The questions dealt with the village, seasonal and annual variations in agriculture, cassava processing and marketing, as well as the diet and the disease konzo which is well known in the area. As almost all the food crops, including cassava, are cultivated by women, they mainly were selected for the groups. One or two men were included in each group since pilot activities showed that this resulted in more open discussions. The dietary information obtained from the focused group interviews was confirmed by interviewing 40 randomly selected households in two villages, using structured questionnaires.

Monthly rainfall data for 1978-88 were obtained from Pater Hoff at Mission Ngondi, 40 km west of the study area.

Results

Population and clinical findings

Demography. The number of inhabitants in each of the 21 villages is given in Fig. 1. The total of 6764 inhabitants (1351 men, 1909 women, 1819 boys, and 1685 girls) in 1568 households gave a mean of 4.3 inhabitants per household. The population consisted of three ethnic groups (Table 1), mostly mixed, in the villages.

Table 1: Prevalence of konzo in the different ethnic groups

<table>
<thead>
<tr>
<th>Ethnic group</th>
<th>Population (total)</th>
<th>No. of patients</th>
<th>Prevalence per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ngongo</td>
<td>4973</td>
<td>74</td>
<td>15*</td>
</tr>
<tr>
<td>Mbala</td>
<td>1255</td>
<td>22</td>
<td>18*</td>
</tr>
<tr>
<td>Hungani</td>
<td>494</td>
<td>14</td>
<td>28*</td>
</tr>
<tr>
<td>Others</td>
<td>42</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>6764</td>
<td>110</td>
<td>16</td>
</tr>
</tbody>
</table>

* No significant difference between the three main ethnic groups with χ²-test (P = 0.08).

Locomotor disabilities. Out of 152 persons with a locomotor disability, 110 fulfilled the criteria for konzo. Sequelae of poliomyelitis were found in 24 persons (4/1000), 12 were crippled due to congenital malformation or had sequelae of trauma, and 6 had unspecified gait difficulties. Besides these 152 persons with locomotor disabilities, another 16 persons spontaneously presented themselves for examination claiming to have had an abrupt onset of minor symptoms similar to konzo which had returned to normal; they showed no observable abnormalities of gait or running but ten had exaggerated knee and/or ankle jerks, and the remaining six showed normal neurological findings. A sister of a konzo-affected woman was obviously simulating a severe spastic gait but lacked exaggerated reflexes.

Clinical findings in konzo patients. Out of the 110 konzo patients, 90% stated that the onset of gait difficulties occurred within one day, and for the remaining 10% within three days. On examination, 19% had a normal gait with a spastic abnormality when running, 45% had a spastic gait but did not use walking sticks, 21% had a spastic gait and regularly used one stick, 4% had a spastic gait and used two sticks, and 11% were unable even to stand. Nine of the 12 bedridden persons also had severe dysarthria. Patellar reflexes were exaggerated in 97% of the patients and not testable in 1% owing to contractures. Ankle reflexes were exaggerated in 79% of the patients and not testable in 8%. Sustainable ankle clonus was present in 43% of the patients, clonus with 3–7 beats in 42%, and was not testable in 8%. Clear plantar responses could only be elicited in 17%.

Blurred vision at the onset was reported in 12% of the patients, but only one had vision complaints during our examination. Five patients had ulcers on
their feet, and two showed lower limb oedema. One of these, a 40-year-old man, had a fullblown kwashiorkor and died some months after the survey. Eleven patients had slightly enlarged thyroid glands but only two were visible when the head was tilted back.

**Occurrence of konzo**

Among the 6764 inhabitants, 110 live, konzo-affected persons were identified (16/1000). Only two small villages were unaffected by the disease (Fig. 1). No statistically significant difference in prevalence was found between ethnic groups (Table 1). A typical history of severe konzo was also obtained for 24 persons who had died 1–7 years after the onset. There were statistically significant higher occurrences of both live and dead patients with konzo in the two largest villages (Table 2), which were inhabited by all three ethnic groups. The sex and age distribution at onset of konzo (Table 3) shows that in this area adult males have a relatively low risk of contracting konzo.

The annual distribution of onsets of konzo in both live and dead patients shows that the disease occurred for the first time ever in the study area in 1974–75 (Fig. 2). Information on earlier onsets were not obtained from records or the staff at Lumbi Health Center and Masi-Manimba hospital. None of the focus groups knew about any earlier onsets in their villages, but old people in 10 of the groups stated that the same paralytic disease occurred in the southern part of the region during colonial times. Five of the groups told us that the disease used to be called "konzo" or "konzi" by former generations in the south. The causes of konzo proposed in the group interviews included lack of food, consumption of wild plants, cold water, insufficiently processed cassava, witchcraft, punishment by God, and hereditary and contagious factors.

The onset of konzo in the 110 live patients occurred mostly during the dry season and in the very beginning of the rainy season, i.e., May to September (Fig. 3). Aggravations of konzo, where a formerly affected individual suffers an abrupt and permanent deterioration of the spastic paraparesis, were documented in 13 of the 110 patients. One of them had suffered 3 such aggravations and all 15 aggravations occurred between May and September. Four were struck by aggravations one year after the first onset, 5 after two years, and the rest after 3–10 years. One woman, who had her first attack in 1986 resulting in permanent walking difficulties, suffered an abrupt aggravation (one month before our examination), which within one day made her unable to stand.

Of the 110 live patients, 36 reported that at least one first-degree relative had konzo, and 8 patients reported that second-degree relatives were affected. The familial clustering was sometimes very pro-

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**Table 2: Distribution of konzo cases, by village size**

<table>
<thead>
<tr>
<th>Konzo patients:</th>
<th>Large (n &gt; 800)</th>
<th>Small (n &lt; 600)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of villages</td>
<td>2</td>
<td>19</td>
<td>21</td>
</tr>
<tr>
<td>Population</td>
<td>1893</td>
<td>4871</td>
<td>6764</td>
</tr>
<tr>
<td>Alive*</td>
<td>53 (29)*</td>
<td>57 (12)</td>
<td>110 (16)</td>
</tr>
<tr>
<td>Dead*</td>
<td>21 (11)</td>
<td>3 (&lt;1)</td>
<td>24 (4)</td>
</tr>
<tr>
<td>Total</td>
<td>74 (39)</td>
<td>60 (12)</td>
<td>134 (20)</td>
</tr>
</tbody>
</table>

* The difference between large and small villages is significant with $\chi^2$-test ($P < 0.001$).

* Figures in parentheses are the number of patients per 1000 inhabitants.

**Table 3: Distribution of konzo patients, according to sex and age at onset**

<table>
<thead>
<tr>
<th>Age at onset (years)</th>
<th>Males</th>
<th>Females</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–2</td>
<td>0 + 0*</td>
<td>0 + 0</td>
<td>0 + 0</td>
</tr>
<tr>
<td>3–14</td>
<td>34 + 3 (2.1)*</td>
<td>24 + 2 (1.6)</td>
<td>58 + 5 (1.8)</td>
</tr>
<tr>
<td>$\geq$15</td>
<td>3 + 5 (0.5)</td>
<td>49 + 14 (2.7)</td>
<td>52 + 19 (1.8)</td>
</tr>
<tr>
<td>Total</td>
<td>37 + 8 (1.2)</td>
<td>73 + 16 (2.0)</td>
<td>110 + 24 (1.6)</td>
</tr>
</tbody>
</table>

* Each set of figures refers to the numbers alive + dead.

* In parentheses are given the estimated mean annual incidence per 1000 inhabitants during 1974–88. Mean population for the period (5894 inhab.) was calculated assuming 2.1% annual population growth, equal in all groups. The estimated numbers of children 0–2 years were subtracted from the number of children 0–15 years.

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*b Expressed as percentage of general price index (20).
nounced, e.g., in one family the father, mother, and both children had all been affected. The father was unable to bear his cruel fate and committed suicide.

**Agricultural aspects**

All the focus groups stated that cassava was the predominant crop, but small quantities of maize were grown in all villages. Groundnuts, squash (*Cucurbita* spp.), bananas, and green leaves (mainly *Amaranthus* spp.) were grown in most villages and the cultivation of sugar cane, yam, sweet potatoes, and rice was reported in fewer than half of the villages. Beans were not cultivated in the area.

Shifting cultivation was mainly practised; the fields were opened by cutting down and burning the vegetation, followed by 2–3 years of cultivation. The fallow periods were for 3 to 7 years, but in 13 out of the 21 villages the groups stated that this had been shortened because of lack of cultivable land. All the fields were located in the river-valley forests.

More than 20 different cassava varieties were reported to be cultivated in the area. Most fields were planted with the bitter varieties that had to be processed prior to consumption. Sweet varieties that could be eaten fresh without processing were only cultivated in small quantities.

Deficient cassava harvests were unknown in the area, the farmers stating that there had never been a poor cassava harvest! However, most villages had experienced poor harvests for other crops, especially groundnuts, which were attributed to delayed rains after the dry season, heavy rains, or invasion by domestic animals, mainly cows. Other domestic animals were goats, sheep, pigs, and poultry.

Water and firewood were available throughout the year, but had to be carried by women from the river valley to the villages, which were often situated on top of the hills with altitude differences of 50–150 metres.

**Cassava processing and toxicity**

The groups stated and we observed that cassava roots were processed into “boulettes” (small balls) and “cossettes” (a French word used for dry cassava roots) in all villages. Both products are pounded into flour which, after sieving, is mixed with hot water to make “luku”, the stiff cassava porridge that constitutes the staple food of the area.

The oldest processing method, used for generations, was to make “boulettes”: the unpeeled roots are first soaked in water for 3–5 days and nights, the wet roots are then peeled and pounded, and the resulting mash is pressed by hand into small balls which are sun-dried for 3–4 days. The processing into “cossettes” was now the main method used: the peeled roots are soaked in water for 3–4 nights, and the entire roots are thereafter sun-dried for 5–7 days.

Cossettes were hardly made at all before 1975. In this year the completion of a new tarmac road dramatically improved the transport facilities from Masi-Manimba zone to the capital Kinshasa (Fig. 1), and thereby the urban demand for staple food rapidly turned cassava into the dominant cash crop. The cassava traders instructed the population to produce cossettes as the fragile balls could not easily be transported. The whole population gradually started to use the cossettes for their own consumption as well, because it was easier to produce and store only one type of product.

To obtain non-toxic cossettes, all groups stated that the roots should be soaked for at least 3 nights, or longer in the dry season when the water is colder. However, 13 of the 17 groups declared that many families frequently soaked the roots for only one or two nights. The reason given was that the need for money forced the women to shorten the processing time. Most groups said that the highest cassava sales occurred in the period May–August and sometimes September–November (Fig. 3); these were the periods when consumption of short-processed roots was most common. The “luku” made from roots soaked for only one night was known by all groups to be elastic and to occasionally cause acute intoxication symptoms within hours of consumption, such as dizziness, fatigue, headache, vomiting, and diarrhoea. We were told that the zone authorities in 1982 had prohibited short-processing owing to the
Table 4: Types of food consumed before the onset of konzo (n = 110)

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Staple:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cassava exclusively</td>
<td>83</td>
<td>0</td>
<td>27</td>
</tr>
<tr>
<td>Short-processed cassava</td>
<td>59</td>
<td>13</td>
<td>38</td>
</tr>
<tr>
<td><strong>Occurrence of acute intoxications after cassava meals</strong></td>
<td>59</td>
<td>22</td>
<td>29</td>
</tr>
<tr>
<td><strong>Supplementary food:</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cassava leaves</td>
<td>83</td>
<td>0</td>
<td>27</td>
</tr>
<tr>
<td>Fish</td>
<td>15</td>
<td>62</td>
<td>33</td>
</tr>
<tr>
<td>Edible wild plants</td>
<td>79</td>
<td>1</td>
<td>31</td>
</tr>
</tbody>
</table>

risk of such acute intoxications. Out of the 40 households interviewed, only one stated that they sometimes soaked the roots for less than two nights, and 37 declared that they sometimes soaked them for less than 3 nights. Acute intoxications after consumption of short-processed cassava roots had been experienced by 15 out of the 40 households, and 59 out of 81 persons affected by konzo declared that they had suffered acute intoxications even before the onset of konzo (Table 4). We noted in all groups and most households that it was a very delicate and sensitive issue to discuss the frequent deviations from the recommended processing practices.

**Diet**

Cassava provided the mainstay of the diet in all villages, according to the focus groups. The roots were normally consumed twice a day as "luku", with supplementary food consisting of a relish of either cassava leaves, other green leaves, mushrooms or fish. Domestic animals were very rarely consumed. The majority of the groups stated that shortage of supplementary food was pronounced during the dry season in June-August and some groups also mentioned shortages in May and September-October (Fig. 3).

Of the 40 randomly selected households, 12, 22 and 23 stated that supplementary-food shortage was common in June, July and August, respectively. Some households also mentioned shortages in April-May, and September-November. One way to cope with the supplementary-food shortage was to eat only one meal a day or to replace one meal by a snack like sweet cassava or bananas. The meals consumed during the 24 hours before our survey in these 40 households are presented in Table 5. In 28% of the households no animal protein was consumed.

Table 5: Survey of 40 randomly selected households concerning meals consumed during last 24 hours

<table>
<thead>
<tr>
<th></th>
<th>Morning</th>
<th>Midday</th>
<th>Evening</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cassava* + fish (+ leaves*)</td>
<td>14</td>
<td>1</td>
<td>22</td>
</tr>
<tr>
<td>Cassava* + meat or egg (+ leaves*)</td>
<td>4</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Cassava* + groundnuts or gourd seeds</td>
<td>3</td>
<td>2</td>
<td>4</td>
</tr>
<tr>
<td>Cassava* + leaves*, mushrooms or fruit</td>
<td>14</td>
<td>6</td>
<td>12</td>
</tr>
<tr>
<td>Yam + nuts</td>
<td>0</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>&quot;Snack meal&quot; (groundnuts or fruit)</td>
<td>4</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Cereals</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Nothing consumed</td>
<td>1</td>
<td>21</td>
<td>2</td>
</tr>
</tbody>
</table>

| Total                    | 40      | 40     | 40      |

* In the form of "luku", as explained in the text.

The persons affected by konzo declared that cassava was the only staple consumed in the period preceding onset of the disease and 80% (62 out of 77) denied eating animal protein during this period (Table 4). The incomplete information from this group is due to absence of relevant informers and the long time before our survey.

**Discussion**

The findings in our neurological examinations are in all relevant parts consistent with those of a neurological study of konzo (5), and our conclusion is that this form of spastic paraparesis in Bandundu is the same disease as the one described in East Africa (6, 7). We also agree that this disease entity is konzo, as described in the first report (1).

**Validity of methods**

The high prevalence, typical onset, specific gait abnormality, and permanent disability, which are characteristic of konzo, facilitated the collection of retrospective epidemiological data by a survey method based on community participation. While the clinical discrimination between konzo and other crippling diseases is precise, that between slightly affected konzo patients and healthy subjects is arbitrary. Our findings suggest that subclinical forms exist and that mildly affected patients may recover, but any significant underestimation of the prevalence of moderate and severely affected patients with konzo is unlikely, for the reasons mentioned above. The diagnosis of konzo in those who died can be regarded as reliable as they were all identified to have suffered from severe forms of the disease.
Cross-checking of the time of onset with the dates on birth certificates of children born at about this time revealed only a few mistakes in the year (within one year's deviation), and estimates of the month of onset varied in a few cases by two months or less.

The focus-group interview method for obtaining information on dietary practices was chosen since we were interested in possible severe and reluctant admissions of deviations from recommended practices and because the reasons for such deviations were unknown to us at the start of the survey. The dietary data obtained in the group interviews are consistent with the results from the household interviews, and the agricultural information from the group interviews are in line with earlier studies (17).

**Epidemiology**

The onset of konzo and its sudden aggravations agree with the earlier findings in Bandundu (1-5), and although almost exclusively limited to the dry season and the first month of the rainy season, the annual incidence varies considerably. As described before (1, 3, 5), adult males and breastfed children have a low risk of contracting konzo. No evidence that ethnic affiliation influences the risk of konzo emerged but familial clustering is common.

No infectious symptoms were reported in relation to the onset and an infectious etiology is unlikely for epidemiological reasons. The estimated mean konzo incidence in the studied population was 2 per 1000 persons per year. The mean incidence of aggravating attacks is estimated at about 20 per 1000 already affected persons per year (15 aggravations in 760 patient-years). Although some milder aggravating attacks may have been missed, there is still a ten times higher risk of konzo attacks among persons formerly affected compared to the general population, which clearly shows that no immunity is acquired. The extensive epidemic outbreaks (1, 6) would indicate a relatively short incubation period but familial clustering, over a period of years, would indicate a long incubation period (5). It is difficult to explain this epidemiological pattern by any known type of neurotropic infectious organism but the pattern is in fact identical with that of other neurological diseases with a dietary etiology, like beriberi (19).

**Evidence for a dietary etiology**

This study of konzo supports an etiology that is linked with exclusive consumption of insufficiently processed cassava roots. It identifies causative mechanisms at community, household and individual levels. The underlying factors at community level are underdevelopment, decline of the agricultural system, and suboptimal market integration. Contributory factors at household level are poverty, high demands on women's time, and no alternative to cassava for cash income, which during the peak sales period induces short-processing of the roots to increase the turnover of sales. The direct causes at individual level are the high dietary cyanide exposure over a period of several days to weeks, a low intake of sulfur-containing amino acids providing substrates for cyanide detoxification (16), and possibly other contributory dietary deficiencies or predisposing factors. This etiology is supported by the following associations.

(1) The appearance of annual konzo outbreaks coincided with the following chain of events: completion of the tarmac road and increased market pressure that turned cassava into the main cash crop from 1974–75 onwards, which considerably shortened the cassava processing time in the area. The reports of acute intoxications after cassava meals following the reduction of processing time indicate high residual levels of cyanogen substances in the cassava flour consumed.

(2) The cyclic variation in the number of annual konzo cases correlates well with the 5-year-long cycles of deflated prices of cossettes in Kinshasa, based on the general price index (20). The highest deflated prices occurred in 1977–79 and 1982–84; almost all onsets occurred in these years (Fig. 2), in this study as well as in an earlier study (5). The reasons given for these price cycles are the farmers' reactions to prices at the planting time (20). Increased planting results in increased supply and subsequent price decreases at harvest 12–24 months later. This, in turn, leads to decreased planting and increased prices two years later, resulting in a cycle of price changes over 4–5 years. The association between high price and high annual konzo incidence suggests that marketing pressure affecting cassava-processing practices is the main link in the chain of factors inducing konzo in Bandundu.

(3) The appearance of new konzo cases during the dry season and start of the rainy season correlates with the worst food supply situation during the year, as stated by both the focus groups and the households interviewed. The supplementary-food shortage during this period results in an extremely monotonous cassava diet which is low in protein, especially of sulfur-containing amino acids. This situation in Bandundu is identical to the dietary situation during konzo epidemics in the lowland (13) and highland (7) areas of East Africa.
(4) The higher incidence of konzo cases in larger villages may be due to the more serious agricultural and dietary crisis in these villages, which is related to the increased pressure on land and the longer distances to get to the fields, with their effects on the women's time and work in order to meet the market demands. The shorter fallow periods and the deterioration of shifting cultivation, leading to a decline in intercrops (17), also aggravates the food supply situation.

(5) The considerably higher frequency of aggravating attacks in already affected subjects is most probably due to deterioration of the diet in the affected families as a consequence of one member, often the mother, being crippled.

(6) The higher incidence among women and children, apart from breastfed infants, is in accord with earlier findings and supports a dietary etiology. Although not verified in the present study, the possibility that men in the families have priority access to protein-rich foods may explain the lower risk in males.

Thus, our findings indicate that konzo is caused by a period of one to several weeks of nearly exclusive consumption of insufficiently processed and therefore still toxic cassava roots. As tropical ataxic neuropathy, described in Nigeria (21), has been associated with several years of moderate toxic exposure from cassava, the difference between the two disease entities may be explained by marked differences in the rate of exposure to the same toxin (7).

Konzo is the main cause of locomotor disability in the studied population. The observed prevalence, 16/1000, exceeds the polio sequelae prevalence, 4/1000, in the studied area as well as the polio sequelae prevalences reported from other tropical countries (22).

Konzo could become a public health problem in other areas that resemble Bandundu, and it is important that medical staff in these areas should be trained to diagnose the disease. The present evidence is strong enough for agricultural and marketing projects to see that efficient cassava processing is carried out in areas of risk. This is essential if this incurable disease is to be prevented.

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Résumé

Zaire: données épidémiologiques en faveur d'une étiologie alimentaire du konzo, une affection du neurone moteur supérieur

L'affection appelée konzo se caractérise par la survenue brutale d'une lésion du neurone moteur supérieur, qui se manifeste par divers degrés de paraparesie spasmodique. Des flambées épidémiques de cette affection ont été décrites au Mozambique, en République-Unie de Tanzanie et au Zaire. Deux hypothèses ont été avancées en ce qui concerne son étiologie: une origine infectieuse au Zaire et une origine alimentaire au Mozambique et en Tanzanie.

Au Zaire, nous avons recherché dans la région de Bandundu, au moyen d'une enquête épidémiologique et d'entretiens portant sur les habitudes alimentaires, un lien éventuel entre la survenue du konzo et la consommation de racines amères de manioc insuffisamment préparées, lien déjà rapporté en Afrique orientale. Sur les 6764 habitants de la région d'étude, on a répertorié 110 malades atteints de konzo et 24 décès pouvant être imputés à cette affection. On n'a observé aucune différence significative de prévalence du konzo selon l'appartenance à l'un quelconque des trois groupes ethniques de la région. Les femmes et les enfants sont les plus touchés. Le manioc amer est un élément dominant de l'alimentation de la population touchée, en particulier pendant la saison sèche, au cours de laquelle la plupart des nouveaux cas de konzo surviennent. Le début, en 1974-1975, des flambées annuelles de Konzo dans cette région a coïncidé avec l'achèvement d'une nouvelle route goudronnée menant à la capitale, qui a facilité le transport du manioc et en a fait la principale culture de rapport. Le commerce intensif du manioc a entraîné un changement des modes de préparation. La méthode habituelle dans cette région consiste à faire tremper les racines pendant trois nuits, afin d'éliminer les glucosides cyanogènes avant consommation. En raison de la rapidité des ventes, ce temps de trempage a été abaissé à 1 à 2 nuits. On sait que le manioc qui n'a trempé qu'une nuit est potentiellement toxique, et la survenue d'intoxications aiguës est l'indice d'une forte consommation de cyanures. Les cas de konzo s'étant déclarés au cours de la saison sèche ont coïncidé avec les plus fortes ventes de manioc de l'année et avec le moment où l'approvisionnement en denrées alimentaires était le plus difficile. La corrélation observée entre les nouveaux cas de konzo et les fluctuations du prix du manioc indique une relation de cause à effet entre les pressions exercées par
le marché, la préparation du manioc, l’exposition aux cyanures et la survenue du konzo. L’aggravation brutale du konzo chez les sujets déjà touchés indique qu’il n’y a aucune immunité acquise.

Ainsi, nos résultats indiquent très nettement que le konzo correspond à une période, qui peut durer de une à plusieurs semaines, où l’on consomme presque exclusivement des racines de manioc n’ayant pas été suffisamment préparées et donc encore toxiques. Cette situation observée au Zaïre, comme celle à laquelle nous avons déjà fait allusion pour les régions écologiquement différentes d’Afrique orientale, est le résultat d’un régime alimentaire riche en cyanures associé à un faible apport en acides aminés contenant des sulfures qui aident à la détoxication des cyanures. Le konzo peut survenir dans d’autres régions présentant les mêmes caractéristiques que celle de Bandundu et il importe que le personnel médical de ces régions soit formé au diagnostic de cette affection. On en sait désormais assez pour faire attention à ce que dans les projets agricoles et commerciaux figurent des dispositions pour que le manioc soit suffisamment préparé dans les régions à risque. Il s’agit là d’un point capital si l’on veut prévenir la survenue de cette maladie incurable.

**References**


