

# Konzo Outbreak, in the South-west of the Democratic Republic of Congo, 1996

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## Summary

**In August 1996, cases of poliomyelitis were reported in Kahemba zone, in the south-west of the Democratic Republic (DR) of Congo. The diagnosis was reviewed and charged to Konzo, a spastic paraparesis attributed to food cyanide intoxication. In order to describe the phenomena, a community-based survey took place and found 237 people affected. The highest prevalence was found in the most isolated part of the zone. The patients suffered from an isolated non-progressive spastic paraparesis of abrupt onset. Children and women were the most affected groups, especially women after childbirth. Most of the patients developed the disease after 1990 with 101 cases in 1996. Cassava processing was the same over time and in all the villages. The study did not fully explain the increased number of cases in 1996 but suggested that complementary investigations regarding micronutrient intakes, especially vitamin A, would be necessary.**

## Introduction

Konzo is a distinct upper motor neurone disease reported from poor rural African communities in the Democratic Republic of Congo (DR Congo),<sup>1-6</sup> Central African Republic,<sup>7</sup> Tanzania<sup>8,9</sup> and Mozambique.<sup>10-14</sup> Konzo is characterized by an abrupt onset of symmetric, isolated permanent and non-progressive spastic paresis. The disease has been attributed to the combined effect of high cyanide and low sulphur intake from an exclusive consumption of insufficiently processed bitter cassava.<sup>11,13,15</sup>

Cassava is a high-yielding root crop, which is one of the main staple foods in the tropics. Bitter cassava varieties are the most commonly used in areas in Africa with poor soil fertility. These varieties contain potentially toxic cyanogenic glucosides which can be eliminated by the disruption of the cell structure allowing an endogenous glycosidase to break down the glucosides. This can be achieved by either mechanical grating or fermentation followed by drying or heating.<sup>16-18</sup> An infectious aetiology<sup>1</sup> has never been confirmed and no retrovirus has been isolated among Konzo patients.<sup>5,7,19</sup> The province of

Bandundu in the DR Congo has been affected several times by epidemics of Konzo. Trolli<sup>20</sup> first documented the disease there in 1938 and out of the 3700 cases documented in Africa, 2000 have occurred in this province.<sup>17</sup>

In August 1996, the Chief Medical Officer reported poliomyelitis cases in the Kahemba region, in Bandundu province in the south-west of the DR Congo, on the border with Angola. After a first investigation, the diagnosis of poliomyelitis was reviewed and changed to Konzo. With the support of Médecins Sans Frontières (MSF), Epicentre carried out an investigation in this region, with the purpose of describing the epidemic and studying factors associated with the recent increase of the disease.

## Materials and Methods

### *Study site*

The city of Kahemba is located in the Bandundu province, 600 km from Kinshasa and 300 km from Kikwit. The Kahemba region has a savannah-type vegetation. The dry season occurs between the months of May and August. Lunda and Tchokwe are the two largest ethnic groups in the region and cassava constitutes their predominant staple food. As a consequence of increasing diamond trade in the region, the population of Kahemba city has multiplied by four in the last 10 years. In 1996 Kahemba city had around 42 000 inhabitants. The city was surrounded by 870 villages which were grouped into 45 locations and four districts with a total of 100 000 inhabitants.

The investigation was conducted in all the villages

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situated along the roads accessible by car or motorbike and covered almost 300 km (Fig. 1). Kahemba city was not included in the study.

### Case finding

In November 1996, two teams visited the region by car or motorbikes when tracks were not suitable for cars (Fig. 1).

For identification of cases we used the WHO definition for Konzo, i.e., visible abnormalities whilst walking or running, abrupt onset in a formerly healthy person and bilaterally exaggerated knee and ankles jerks without signs of spinal disease.<sup>17</sup> Active case finding of persons with locomotor disabilities was conducted with the assistance of village and religious leaders. All subjects were examined by a senior physician and classified as a case of Konzo if they fitted the WHO diagnostic criteria.

For each case identified, demographic data (sex, age, ethnic group, residence) and the history of the disease were collected on a standardized questionnaire. The date of onset was estimated with the help of a local event calendar. The gait difficulty was assessed whilst running and walking. We used the WHO classification to categorize the disability as mild when the patient did not require a regular

walking aid, as moderate when he used sticks, and as severe when he was bedridden or unable to walk without support from another person. The patient's complaints of visual problems were also recorded.

Prevalence and incidence rates were calculated using data collected by the local administration in 1995 as the denominator population. The sex and age distributions were estimated from the 1985 census.

### Group discussion

In the villages in which Konzo patients were identified, information on food crops, dietary habits, and cassava processing, as well as knowledge and attitudes on Konzo, was obtained through group discussions with the residents.

## Results

### Case finding

In November 1996, the two teams visited 39 of the 45 existing locations (86.7 per cent), the other six locations were not accessible. They collected information on 441 people who presented locomotor disability. Of these, 174 were absent from their homes and could not be examined. Of the remaining 267 persons, a total of 237 met the case definition. The 30 subjects excluded had either a non-symmetrical illness, a slow onset of symptoms (more than 1 week), or normal walking and running.

### Clinical description of cases

Among the 237 patients meeting the case definition, the illness was considered mild for 166 (70 per cent), moderate for 59 (25 per cent), and severe for 12 (5 per cent). Out of the 166 persons walking without help, 25 (15 per cent) exhibited problems in their gait when running but had normal walking.

From the 233 patients for whom information was available, 122 (52 per cent) reported an onset of the symptoms in less than 24 h. Almost all patients (223/230) presented with an ankle clonus (Table 1).

### Epidemiological description

Konzo patients were identified in 19 of the 39 (49 per cent) locations visited. Overall disease prevalence was 2.4 per 1000 inhabitants but varied widely from one location to another (range 0.2 to 31.7 per 1000 with a median at 1.8 per 1000). The highest prevalence of Konzo per location was observed around Tshifamesu and Mwa-mushiko (Fig. 1).

The year of onset was available for 234 patients. Of these, 191 (82 per cent) had developed the disease after 1990, and 101 (43 per cent) during 1996. An old man reported having contracted Konzo in 1925. This information was checked with local events. The next case occurred in 1960.

Among the 237 patients, 39 (16 per cent) had experienced two attacks of Konzo. The month of

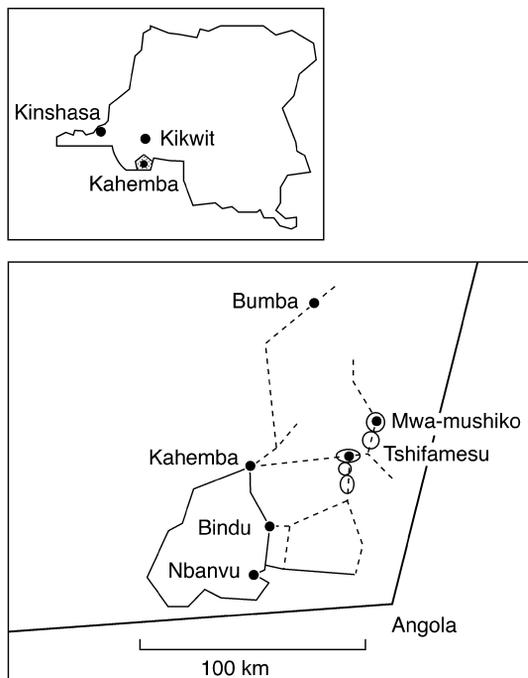


FIG. 1. Democratic Republic of Congo and Kahemba zone. —, Access by car; ---, access by motorbike; ○, location with prevalence > 15/1000.

TABLE 1  
Distribution of clinical symptoms among patients  
with Konzo in Kahemba zone, DR Congo,  
November 1996

Clinical findings	Frequency	%
Ankle clonus	223/230	97
Onset in less than 24 h	122/233	52
Dysarthria noticed during the examination	78/233	34
Exaggerated reflex on the upper limbs	37/222	17
Babinski	26/220	12
Complaints of visual troubles	24/233	10

onset was available for 163 people who reported their first attack after 1990 and amongst them, 128 (79 per cent) were affected during the dry season (Fig. 2).

The male : female ratio was 0.7. For 151 people (66 per cent), the first onset of the disease occurred before they were 16 years old and in this group the sex ratio was 1. For those affected when adults, the male : female ratio was 0.2. No cases were reported amongst children less than 2 years old. The ethnic group was known for 229 people of whom 160 (68 per cent) were Tchokwe and 69 (29 per cent) Lunda. We

did not have the population distribution by ethnic group. Out of the 143 female patients, 65 contracted Konzo whilst of reproductive age. Detailed information was available for 37 of them. All but one of them developed the disease within a year after delivery. Of these, 22 (59 per cent) developed the disease in the first 3 months after delivery. Only one developed symptoms during pregnancy.

Adult females had a prevalence three times higher than adult males who had the lowest (Table 2). Among children, the prevalence was not statistically different.

#### Focus group discussions on cassava processing

Cassava roots were usually soaked for 2 nights (one more night during the dry season), then peeled, sundried for between 2 (if roots squeezed into balls) to 7 days (if cassava as whole roots) and finally, pounded into flour. The flour was mixed with water to make a stiff porridge called *fufu*. No maize was added to this porridge. The relish eaten with the *fufu* was usually prepared with green leaves although during the dry season, other food was scarce and the *fufu* was often eaten by itself. No change was reported in cassava processing over time and the same variety of cassava was cultivated. However, many people complained of decreasing yields of root crops over the last

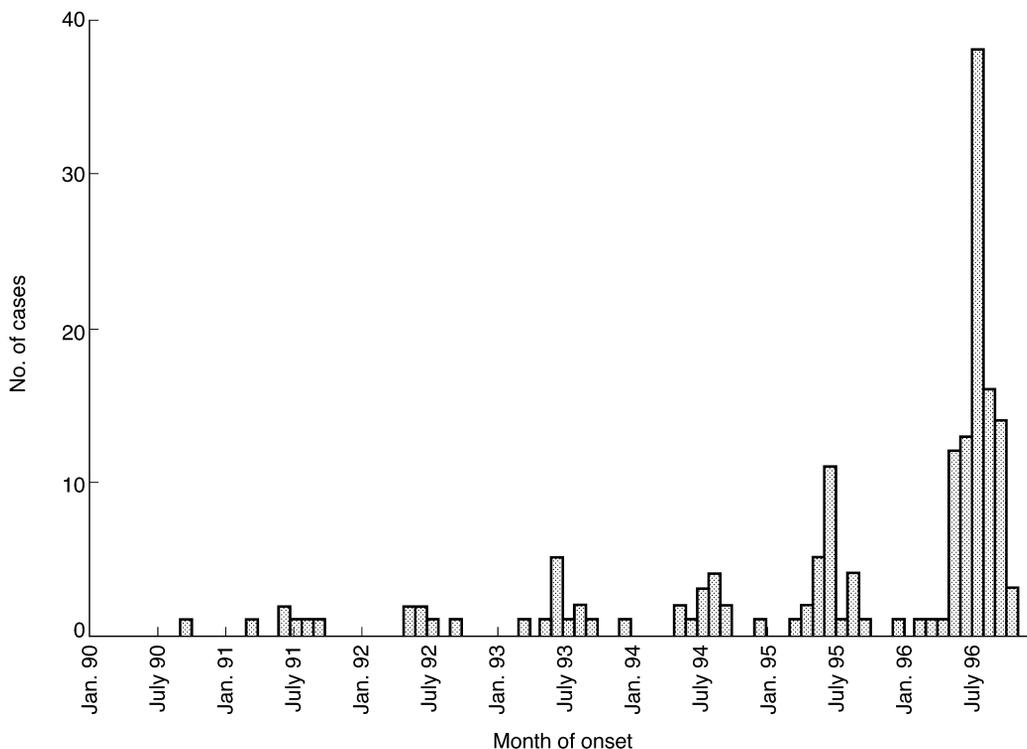


FIG. 2. Month of onset of Konzo patients ( $n = 163$ ) from January 1990 to October 1996, Kahemba zone, DR Congo.

TABLE 2

Number of Konzo patients by sex and age group, estimated prevalence per 1000 inhabitants and relative risk, Kahemba zone, DR Congo, 1996

	Cases	Prevalence	Relative risk	95% CI
Male adult	22	1.2	Reference	
Female < 16 years	61	2.0	1.7	1.1–2.8
Male < 16 years	72	2.4	2.0	1.3–3.3
Female adult	82	3.8	3.2	2.0–5.1
Total	237	2.4		

decade. There were no reports of droughts although this could not be confirmed, as no rainfall data was available.

The information concerning cassava processing, diet, climate or cassava crops did not differ between the locations visited.

### Discussion

This investigation confirmed that Konzo was endemic and highly prevalent in the Kahemba region, although the lack of access to many areas will have underestimated the total number of cases.

An increased number of cases during 1996 suggested that an outbreak of Konzo affected this region. This is one of the largest outbreaks reported in the literature, surpassed by those reported by Trolli in DR Congo in 1938<sup>20</sup> and the two outbreaks in Mozambique.<sup>10,14</sup>

The disease mainly affected those living in isolated locations with no access by car, women in the reproductive age group and children over 2 years old. The number of Konzo cases was higher during the dry season. The case finding did not include the people who died with the disease and thus one must interpret the epidemic curve with caution. Nevertheless, these former characteristics have been frequently described in other Konzo epidemics.<sup>1–3,8–10,17</sup>

This study showed a high proportion of women affected within the first term following infant delivery. Increased protein and vitamin A requirements during pregnancy, lactation and child growth could explain why women and children are at an increased risk of contracting Konzo. It has been demonstrated that serum levels of vitamin A following delivery are low compared to non-pregnant women,<sup>23</sup> but then requirement of vitamin A seems to decrease as retinol levels in breastmilk decline progressively during breastfeeding.<sup>24,25</sup> This could explain the high percentage of women affected at an early stage after delivery. Indeed, vitamin A has been given to animals as an additive for cassava-based diets in order to overcome the effects of cyanogenic glucoside.<sup>26</sup>

During the study, we tried to correlate the annual number of cases of Konzo and the price of the palm oil, rich in vitamin A, but we were unable to obtain the necessary information. At the time, the local economy seemed destabilized by the diamond trade and American dollars were used in the market of Kahemba city. Eventually, this could have resulted in an increase in the price of the palm oil and could explain the distribution of disease among the populations who were poorest and furthest from the supply routes (e.g. Kahemba city and the main roads).

Konzo is a major public health problem in Kahemba region. In the affected communities, the social consequences of the disease were dramatic. In the visited villages, the affected women could not farm and thus could not feed their families. Many of their husbands had already left them and only a few women received any assistance from relatives. Others were living on the edge of the village, abandoned by the community and suffering from severe malnutrition. The majority of these women, if not all, knew that they were sentenced to death. Unfortunately, it could be assumed that, once grown, affected girls would suffer a similar fate although children with Konzo seemed to be well integrated in the community. The social consequences strengthen the need for further quick investigations in order to implement appropriate control measures.

The reason why Konzo is increasingly affecting isolated areas in the Kahemba region since 1990 is still unclear. The interviews or the focus group discussions did not identify any changes in cassava processing or in the climate that could explain a mono-consumption of insufficiently processed cassava. Konzo seems to affect particularly the population of Bandundu. In this province, cassava flour is not mixed with maize as it is done in the rest of the country (maize increases the level of sulphur that is needed to detoxify the cyanide ingested). Therefore, the economic crisis that affected the country for several years has probably had a major impact in this particular region and led to an increase of Konzo over time.

The socioeconomic burden on this impoverished region is heavy. In order to reduce this burden, preventive measures based on information and educational activities, as well as physical rehabilitation programmes, need to be planned and implemented. The role of nutritional deficiencies need to be studied further, especially the role of vitamin A, as they may be one of the cornerstones of any programme aiming to control the disease.

## References

- Carton H, Kayembe K, Kabeya O, *et al.* Epidemic spastic paraparesis in Bandundu (Zaire). *J Neurol Neurosurg Psychiatr* 1986; 49: 620–27.
- Tylleskar T, Banea M, Bikangi N, Fresco L, Persson LA, Rosling H. Epidemiological evidence from Zaire for a dietary aetiology of Konzo, an upper motor neurone disease. *Bull WHO* 1991; 69: 581–89.
- Banea M, Bikangi N, Nahimana G, Nunga M, Tylleskar T, Rosling H. Haute prévalence de Konzo associée à une crise agro-alimentaire dans la région du Bandundu au Zaïre. *Ann Soc Belge Med Trop* 1992; 72: 295–309.
- Tylleskar T, Banea M, Bikangi N, Cooke R, Poulter N, Rosling H. Cassava cyanogens and Konzo, an upper motoneuron disease found in Africa. *Lancet* 1992; 339: 208–11.
- Tylleskar T. L'étiologie du konzo. Etudes sur une affection paralytique en Afrique. Thèse de la Faculté de Médecine, Uppsala, Sweden, 1994.
- Banea M, Tylleskar T, Rosling H. Konzo and Ebola in Bandundu region of Zaire. *Lancet* 1997; 349: 621.
- Tylleskar T, Legue FD, Peterson S, Kpizingui E, Stecker P. Konzo in the Central African Republic. *Neurology* 1994; 959–61.
- Howlett P, Brubaker GR, Mlingi N, *et al.* Konzo, an epidemic upper motor neuron disease studied in Tanzania. *Brain* 1990; 113: 223–35.
- Howlett P, Brubaker G, Mlingi N, Rosling H. A geographical cluster of Konzo in Tanzania. *J Trop Geograph Neurol* 1992; 2: 102–8.
- Ministry of Health, Mozambique. Mantakassa: an epidemic of spastic paraparesis associated with chronic cyanide intoxication in a cassava staple area of Mozambique. 1. Epidemiology and clinical and laboratory finding in patients. *Bull WGO* 1984; 62: 477–84.
- Ministry of Health, Mozambique. Mantakassa: an epidemic of spastic paraparesis associated with chronic cyanide intoxication in a cassava staple area of Mozambique. 2. Nutritional factors and hydrocyanic acid content of cassava products. *Bull WHO* 1984; 62: 485–92.
- Essers AJA, Alsen P, Rosling H. Insufficient processing of cassava induced acute intoxications and the paralytic disease Konzo in a rural area of Mozambique. *Ecol of Food Nutr* 1992; 27: 17–27.
- Cliff J, Lundqvist P, Martensson J, *et al.* Association of high cyanide and low sulphur intake in cassava-induced spastic paraparesis. *Lancet* 1985; 30: 1211–13.
- Cliff J, Nicala D, Sauter F, *et al.* Konzo associated with war in Mozambique. *Trop Med Int Health* 1997; 2: 1068–74.
- Tylleskar T, Banea M, Bikangi N, *et al.* Dietary determinants of a non-progressive spastic paraparesis (Konzo): a case-referent study in a high incidence area of Zaire. *Int J Epidemiol* 1995; 24: 949–56.
- Banea M, Poulter N, Rosling H. Shortcuts in cassava processing, and risk of dietary cyanide exposure in Zaire. *Food Nutr Bull* 1992; 14: 137–43.
- World Health Organization. Konzo: a distinct type of upper motoneuron disease. *Weekly Epidemiol Rep* 1996; 71: 225–32.
- Hovette P, Molinier S, Debonne JM, *et al.* Le manioc et sa pathologie. *Med d'Afrique Noire* 1992; 39: 204–6.
- Tylleskar T, Banea M, Bottiger B, Thorstensen R, Biberfeld G, Rosling H. Konzo, an epidemic spastic paraparesis in Africa, is not associated with antibodies to HTLV-I, HIV, or HIV gag-encoded proteins. *Letters to the Editor. J Acquir Immune Defic Syndr Hum Retrov* 1996; 12: 317–19.
- Trolli G. Résumé des observations réunies au Kwango, au sujet de deux affections d'origine indéterminée. Brussels: Fonds Reine Elisabeth, 1938; 1–36.
- Dibley MJ, Goldsby JB, Staehling NW. Development of normalized curves for the international growth reference: historical and technical considerations. *Am J Clin Nutr* 1987; 46: 736–48.
- Dean AJ, Coulombier D, Smith CD, *et al.* Epi Info, Version 6.04: a word-processing, database, and statistics program for epidemiology on microcomputers. Centers for Disease Control and Prevention, Atlanta, Georgia, 1996.
- Dostalova L. Vitamin status during purperium and lactation. *Ann Nutr Metab* 1984; 28: 385–408.
- Lauber E, Reinhardt M. Studies on the quality of breast milk during 23 months of lactation in a rural community of the Ivory Coast. *Am J Clin Nutr* 1979; 32: 1159–73.
- Sagarmay B, Barua S, Tarannum S, Nahar L, *et al.* Retinol and alpha-tocopherol content in breast milk of Bangladeshi mothers under lower socio-economic status. *Int J Food Sci Nutr* 1997; 48: 13–18.
- Kamalu, BP. The adverse effects of long-term cassava consumption. *Int J Food Sci Nutr* 1995; 46: 65–93.