

The care of the limb deficient child in Japan

S. KAKURAI* and M. KIDA†

* *Teikyo University Ichihara Hospital, Ichihara City, Japan*

† *Teikyo University School of Medicine, Ichihara City, Japan*

Abstract

The incidence as well as the current state of the management of the limb deficient child in Japan is described.

General medical and welfare services in Japan

The medical service system in Japan is well organized and includes the diagnosis, treatment and follow-up care of limb deficient children. Once the child has been diagnosed as physically disabled due to limb deficiencies and/or congenital malformations, he or she has the right to receive welfare assistance under the Law for the Welfare of Physically Disabled Children. They are registered and issued with a handbook for identification and access to services. The welfare offices are in charge of the manufacture and distribution, as well as the repair, of the prosthetic appliances. Unfortunately, electric arm prostheses are not included in the inventory at the present time. Those children for whom surgical conversion seems appropriate are evaluated for short-term hospitalization at the medical agencies designated by the Ministry of Health and Welfare. However, not every facility is experienced in the management of such children, as will be shown later in the results of a questionnaire survey.

Incidence of limb deficiencies present at birth and activities of parents organizations in Japan

Thalidomide embryopathies were prevalent during the early 1960's in Japan and 306 patients (171 males and 135 females) were registered (Kida, 1987).

All correspondence to be addressed to S. Kakurai, Department Physical Medicine and Rehabilitation, Teikyo University Ichihara Hospital, 3426-3, Anegasaki, Ichihara City 299-01, Japan.

A survey has been conducted by the Japan Physician's Association for Maternal Welfare (JAMW) for monitoring birth defect babies since 1982. Approximately 120,000 new-born babies have been examined annually at 250 gynaecological hospitals and facilities throughout the country and the results registered with the International Clearing House for Birth Defects. Monitoring System (ICBDMS). According to the statistical study of Kida (1989), new-born children with limb-reduction deformity are born at a rate of 7 per 10,000 births; therefore, approximately 1,000 affected infants are born annually. Table 1 shows the frequency of limb-reduction deformities recorded by the JAMW between 1978 and 1986.

The Association of Parents of Children with Congenital Malformation of the Limbs (APCCML) was organized in 1979 and more than 1,000 families are joint members. Since the establishment of the association one of the authors has been engaged in 66 screening surveys throughout the country and has examined a total of 1,441 cases (Kida, 1989). Table 2 shows a comparison of results of limb deficiencies present at birth registered by the JAMW and screening surveys done at the APCCML. The APCCML have many activities for the psychological support of its members and also to encourage the development of social understanding.

Apart from this study, the authors wanted to investigate the situation of congenital limb deficient children who had been hospitalized in facilities for crippled children and at children's hospitals throughout the country. Some 79 questionnaires were sent and 49 answers received (62% response ratio); 23 facilities had never experienced such cases during the past

Table 1. Frequency of limb reduction deformities registered by the JAMW (1978-86) () indicates per 10,000 births

	1978-81	1982-86	Total	%
Oligodactyly/finger	94 (1.70)	128 (1.92)	222 (1.82)	28.1
Oligodactyly/toe	53 (0.96)	50 (0.75)	103 (0.84)	13.0
Hand defect	14 (0.25)	—	14 (0.25)	1.8
Split hand	—	19 (0.29)	19 (0.29)	2.4
Split foot	—	9 (0.14)	9 (0.14)	1.1
Phocomelia	10 (0.18)	10 (0.15)	20 (0.16)	2.5
Transverse amelia/upper limb	—	20 (0.30)	20 (0.30)	2.5
Transverse amelia/lower limb	—	18 (0.27)	18 (0.27)	2.3
Hypoplasia/upper limb	94 (1.70)	85 (1.28)	179 (1.47)	22.6
Hypoplasia/lower limb	98 (1.77)	82 (1.23)	180 (1.48)	22.8
Constriction band syndrome	—	7 (0.10)	7 (0.10)	0.5
Subtotal	363	428	791	100.0
Total births (JAMW)	553,167	665,861	1,219,028	
Incidence of limb Reduction deformity (per 10,000 births)	6.562	6.427	6.488	
Total births (whole country)	6,457,567	7,328,382	13,785,949	
Estimated number of limb reduction deformities	4,237	4,710	8,944 [993·8/yr]	

Table 2. Comparison of results of limb reduction deformities registered by the JAMW and screening surveys done at the APCCML (Kida, 1989)

JAMW	%	APCCML Surveys	%
Oligodactyly/finger	28.1	Oligodactyly/finger and toe	3.9
Oligodactyly/toe	13.0		
Hypoplasia/upper limb	22.6	Hypoplasia/upper and lower limb	30.4
Hypoplasia/lower limb	22.8	Ulnar aplasia	1.9
Hand defect	1.8	Poland Synbrachydactyly	8.2
		Syndactyly/finger and toe	4.8
		Brachydactyly	1.9
		Polydactyly	1.5
Split hand	2.4	Split hand and foot	9.7
Split foot	1.1		
Phocomelia	2.5	Phocomelia	0.5
Transverse amelia/upper limb	2.5	Transverse amelia/upper and lower limb	1.9
Transverse amelia/lower limb	2.3		
Constriction band syndrome	0.5	Constriction band syndrome	26.6
		Others: Congenital multiple arthrogryposis	1.9
		Microtia	6.8
		[Total 253 Patients]	

Table 3. Number of cases experienced in 26 facilities for crippled children and children's hospitals between 1988-1990

Type of Case	Number of Cases	Percentage
Unilateral upper limb	54	32.1
Bilateral upper limb	19	11.3
Unilateral lower limb	61	36.3
Bilateral lower limb	21	12.5
Unilateral upper and lower limb	7	4.2
Triple limb	3	1.8
Quadruple limb	3	1.8
Total	168	100

Table 4. Number of cases presented at childrens hospitals between 1988-1990 showing the number of surgical procedures performed and the number of prostheses supplied

Category	Unilateral cases	Bilateral cases	Total	Number of cases with surgical procedures	Number of cases fitted with prostheses
Upper limbs					
<i>Transverse deficiencies:</i>					
Shoulder	13	8	21	0	11 (52.4%)
Upper and forearm	17	2	19	0	12 (63.2%)
<i>Longitudinal deficiencies:</i>					
Complete	17	9	26	5 (19.2%)	7 (26.9%)
Partial	29	6	35	1 (2.9%)	2 (5.7%)
Total	76	25	101	6 (5.9%)	32 (31.7%)
Lower limbs					
<i>Transverse deficiencies:</i>					
Hip	0	2	2	0	1 (50.0%)
AK and BK	10	3	13	10 (76.9%)	12 (92.3%)
<i>Longitudinal deficiencies:</i>					
PFFD	7	3	10	4 (40%)	6 (60%)
Tibial:					
Complete	7	7	14	9 (64.3%)	14 (100%)
Partial	13	1	14	14 (100%)	6 (42.8%)
Fibular:					
Complete	16	3	19	6 (31.6%)	11 (57.9%)
Partial	2	0	2	0	1 (50%)
Phocomelia*	10	3	13	1 (7.1%)	12 (85.7%)
Total	65	22	87	44 (50.6%)	63 (72.4%)

* Precise classification unknown.

three years, the remaining 26 facilities had experienced a total of 168 cases (Table 3). Table 4 shows the number of cases seen, the number of surgical procedures performed and the number of cases who have been fitted with prostheses.

Fundamental treatment method for the upper limb deficient child

Among unilateral upper limb amputees in Japan — a country whose people are considered to be generally uniform in appearance and where many value cosmesis over function — most including the parents of limb deficient children, want a cosmetic hand. The split hook is seldom prescribed and used for unilateral cases because of cosmesis (Kakurai *et al.*, 1989). Conversely, bilateral cases are fitted and trained with functional hooks. Due to the small experience with externally powered arm prostheses for higher levels of amputation in this country, most amelic cases have never been fitted with functional arms or externally powered arm prostheses. However, they have been trained to be independent in activities of daily living using their feet.

Fundamental treatment method for the lower limb deficient child

The fundamental programmes for lower limb deficient children in Japan are similar to those of other advanced countries. Teenage amputees are fitted with endoskeletal lower limb prostheses; the diameter of the tubes for such prostheses is 25 mm, whereas that for adults is 30 mm. Although many adult amputees have benefited from turntables and torque absorbers in order to adapt to Japanese domestic life styles, those for youngsters are unfortunately not available at the present time.

Surgical conversion of congenital lower limb deficiencies in Japan

In order to determine the opinions of orthopaedic surgeons engaged in the treatment of crippled children, a questionnaire survey of surgical conversions was sent to them. The following are the results among these doctors of optional treatment methods for typical unilateral congenital lower limb deficiencies; the numbers in the parentheses indicate the number of responses.

Unilateral PFFD (Aitken-Type C):

Knee arthrodesis and Syme's amputation (4); Van Nes rotation osteotomy of the leg (3); hip surgery and leg lengthening (2); leg lengthening (2); hip surgery, knee arthrodesis and Syme's amputation (1); conservative treatment (3).

As the incidence of PFFD in Japan is rather low, the opinions expressed are not statistically significant.

Unilateral total tibial deficiency

Knee disarticulation (7); Brown procedure (5); Brown procedure or knee disarticulation (2); Syme's amputation or knee disarticulation (1); leg lengthening (1); below-knee amputation (1); conservative treatment (5).

It seems from the authors' experience that absolute indications for the Brown procedure are not very extensive.

Unilateral total fibular deficiency

Syme's amputation (9); leg lengthening (2); Syme's amputation or leg lengthening (1); conservative treatment (14).

Although the answers are few in number, these opinions reflect the Japanese doctors' philosophy of surgical conversion of typical lower limb-deficiencies.

Education curriculum for medical and allied health professions on the management of the limb deficient child

That part of the curriculum dealing with the management of limb-deficiency is generally unsatisfactory, with the exception of a one week course on prosthetics and orthotics held under the auspices of the Ministry of Health and Welfare. Since 1975 more than 2,000 doctors have attended this course. At the present time, only one hour of lectures by the authors is included.

Special prostheses and devices for the limb deficient child developed in Japan

Original prostheses for limb deficient children developed in Japan are few, except for the passive hand for small children (Fig. 1), special socket design for unilateral hemipelvectomy developed by R. Takahashi in which the body weight is supported by the sound ischium (Fig. 2) and a torque absorber made of titanium alloy (Betto *et al.*, 1989) (Fig. 3).

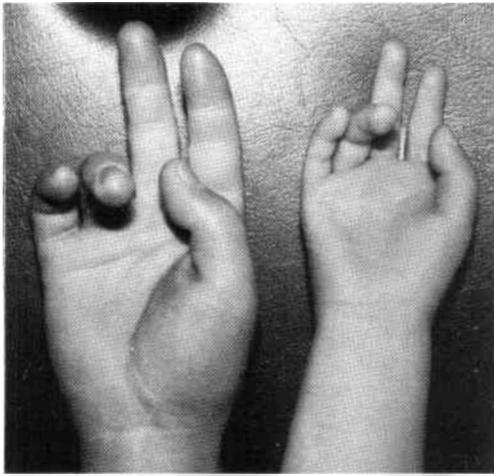


Fig. 1. Passive hand for unilateral upper limb amputated adults (left) and small children (right)

Nowadays most limb deficient children, including multiple cases, attend normal public schools. The curriculum in music of normal lower primary schools, from the third grade until the completion of compulsory education, necessitates that every child must play the recorder-flute. This is very difficult and sometimes impossible for the upper limb deficient and/or handicapped child. This has led us to the development of two kinds of recorder-flute for the unilateral arm amputee or handicapped child (Kakurai *et al.*, 1983). The first type for unilateral upper limb amputees uses a special mechanism on the outside of the flute to cover each hole. This can be manipulated by the child's sound fingers (Fig. 4). The second type, for those who have more than one finger, such as a cleft hand, covers the original holes and opens new holes so that the

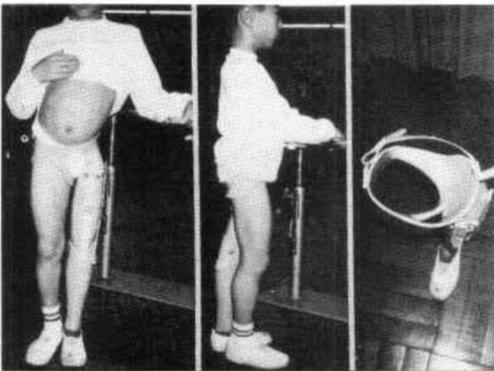


Fig. 2. Special socket design for unilateral hemipelvectomy (R. Takahashi).



Fig. 3. Unilateral PFFD at two years old, fitted with extension brace and at 12 years old Sime's amputation was performed and the child was fitted with endoskeletal modular prosthesis with torque absorber made of titanium alloy.



Fig. 4. Special recorder-flute for the unilateral amputated child, the amputated side is fitted with Swedish myoelectric hand.

remaining digit can operate it quite easily. Since 1975, thanks to the special musical instruments, several thousand children have been able to play the recorder-flute with their peers.

Although the medical and social welfare problems of geriatrics have been overwhelming in recent years in this country, more effort should be continuously directed towards the welfare of limb deficient children.

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