

# Turned head – adducted hip – truncal curvature syndrome

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## Abstract

**One hundred and eight neonates and infants who showed the clinical triad of a head turned to one side, adduction contracture of the hip joint on the occipital side of the turned head, and truncal curvature, which we named TAC syndrome, were studied. These cases included seven with congenital and five with late infantile dislocations of the hip joint and 14 who developed muscular torticollis. Forty one were among 7103 neonates examined by one of the authors. An epidemiological analysis confirmed the aetiology of the syndrome to be environmental. The side to which the head was turned and that of the adducted hip contracture showed a high correlation with the side of the maternal spine on which the fetus had been lying. TAC syndrome is an important asymmetrical deformity that should be kept in mind during neonatal examination, and may be aetiologically related to the unilateral dislocation of the hip joint, torticollis, and infantile scoliosis which develop after a vertex presentation.**

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It has been postulated that abnormal intra-uterine pressure produces many congenital postural deformities.<sup>1-5</sup> Mau reported a clinical combination of congenital deformities that he referred to as the 'seven signs syndrome',<sup>6</sup> and Lloyd-Roberts and Pilcher described the 'moulded baby syndrome' consisting of a tetrad of postural scoliosis, skull moulding, rib cage moulding, and pelvic obliquity.<sup>7</sup> The ipsilateral coexistence of dislocation of the hip joint with a palpable sternomastoid mass<sup>8-10</sup> or with plagiocephaly<sup>11 12</sup> suggests a specific intrauterine asymmetrical posture of the fetus. Most of these reported cases, however, were not in neonates but rather in older infants, and several postnatal factors, such as the development of a sternomastoid tumour or persistently turned head, trunk and pelvis to one side, especially in the supine position could clearly have modified the clinical signs. We observed a group of neonates who showed a characteristic set of asymmetrical postural deformities identified during the routine newborn examination. Their three common signs were a strong tendency to turn the head to one side, adduction contracture of the hip joint or restriction of hip abduction in flexion correlating with the occipital side of the turned head, and curvature of the trunk. We termed this combination of deformities the TAC syndrome, for turning, adduction, and

curvature. An epidemiological analysis was carried out to determine whether the intra-uterine environment of these asymmetrically deformed babies had been restricted. The clinical course of each feature of the clinical triad was also analysed to determine whether TAC syndrome is aetiologically related to any subsequent paediatric disorders.

## Patients and methods

We studied a total of 108 cases with TAC syndrome. Of them, 41 were among a total number of 7103 neonates personally examined by one of the authors (CH) at newborn examinations conducted at hospitals in four cities since 1981. Thirteen were referred neonates. The remaining 54 were among infants aged from 10 days to 3 months who were referred to one of the authors (CH) with suspected torticollis, congenital dislocation of the hip joint, truncal curvature or leg length discrepancy. The mothers of the infants in the latter group stated that they showed a strong tendency to turn the head to one side and that this tendency was apparent immediately after birth. The epidemiological survey for the 54 neonates was conducted separately from that for the 54 infants. Control data were obtained from the birth records of 518 normal live babies examined by one of the authors (CH) in 1982, in Moriyama City, Japan. The clinical course of each of the triad of signs was followed up until the contractures and asymmetry disappeared, or for up to seven years in those patients with dislocation of the hip joint and scoliosis.

## Results

All 108 patients showed the clinical triad described above (fig 1). Statistical data for the neonates, infants, and all patients are shown in table 1. The neonatal incidence was 5.8/1000 live births (41/7103). There was a significant preponderance of girls, first born children (neonates and total), those born in the winter months (neonatal and total), those born with lower birth weight (infants and total), those whose mothers had small abdominal circumference (neonates and total), and those whose mothers had their first child at higher than the average age (neonates and total). One hundred and two babies were born by vertex presentation, and the remaining six by breech presentation. In three of the latter, the mother had a uterus bicornis. Changes in the presentation or position of the fetus in the uterus during the last 10 weeks of gestation were noted by examining the weekly

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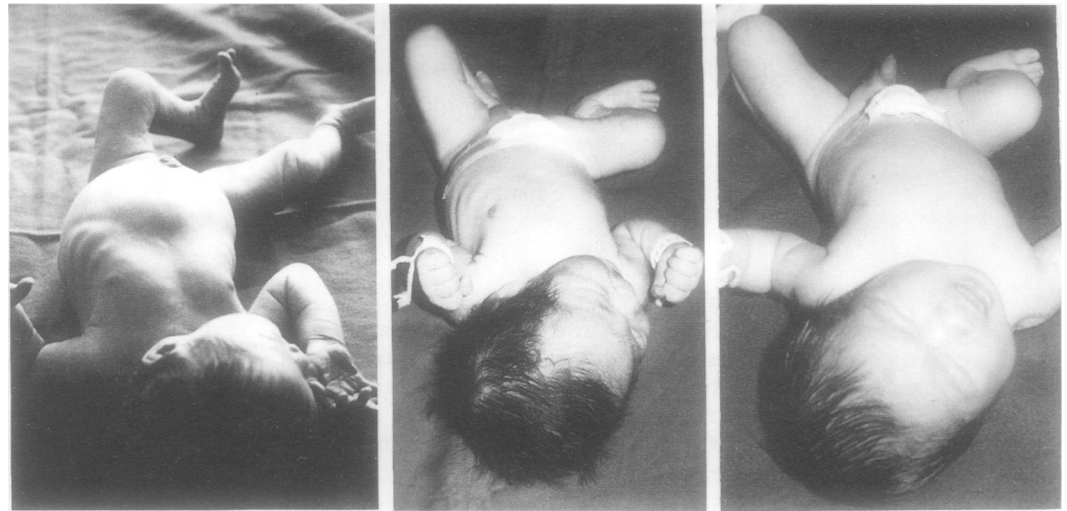


Figure 1 Clinical appearance of neonates with TAC syndrome. These three babies are turning their heads to the right side, and showing adduction contracture of the left hip on the occipital side. The hip joint on the right (facial) side is markedly abducted.

Table 1 The epidemiological findings for the patients with TAC syndrome

	Neonatal (n=54)	Infantile (n=54)	Total (n=108)	Controls (n=518)
Female/male	41/13**	38/16**	79/29***	267/251
First born/others	36/9***	25/25	61/34***	201/317
Winter/summer births	34/20*	30/24	64/44*	248/270
Birth weight (g)†	3028 (409)	2897 (448)**	2953 (433)**	3121 (390)
Mother's abdominal circumference (cm)†	88.1 (2.7)**	89.3 (4.5)	88.8 (4.0)*	90.3 (5.4)
Maternal age of first born (years)†	28.2 (4.0)**	25.4 (2.4)	27.1 (3.7)*	25.8 (3.0)
Change in fetal position (times/10 weeks)†‡	1.04 (1.2)***	0.69 (1.0)***	0.78 (1.07)***	2.33 (1.69)

\*p<0.05, \*\*p<0.01, \*\*\*p<0.001. †Mean (SD); ‡During the last 10 weeks of pregnancy.

obstetric outpatient records. The frequency of change of position in utero was significantly lower than that in the normal fetuses. When the fetus' back was on, for example, the left side of the mother, that is, the left side of the fetus in vertex presentation rested on the mother's spine (fig 2), the adduction contracture of the hip joint and muscular torticollis were almost always on the patient's left side. Similarly, when the fetus' back was on the maternal right side, these signs were almost always on the right; those correlations were highly significant (table 2). The incidence of prematurity, postmaturity, oligohydramnios, and twin births did not differ significantly from that in the control group. A familial survey was conducted on 62 probands. Of the 39 siblings, 17 (44%) had asymmetrical

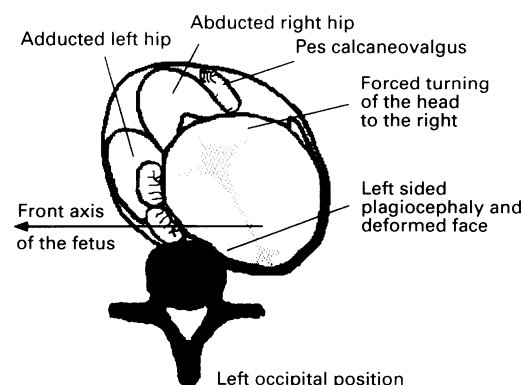


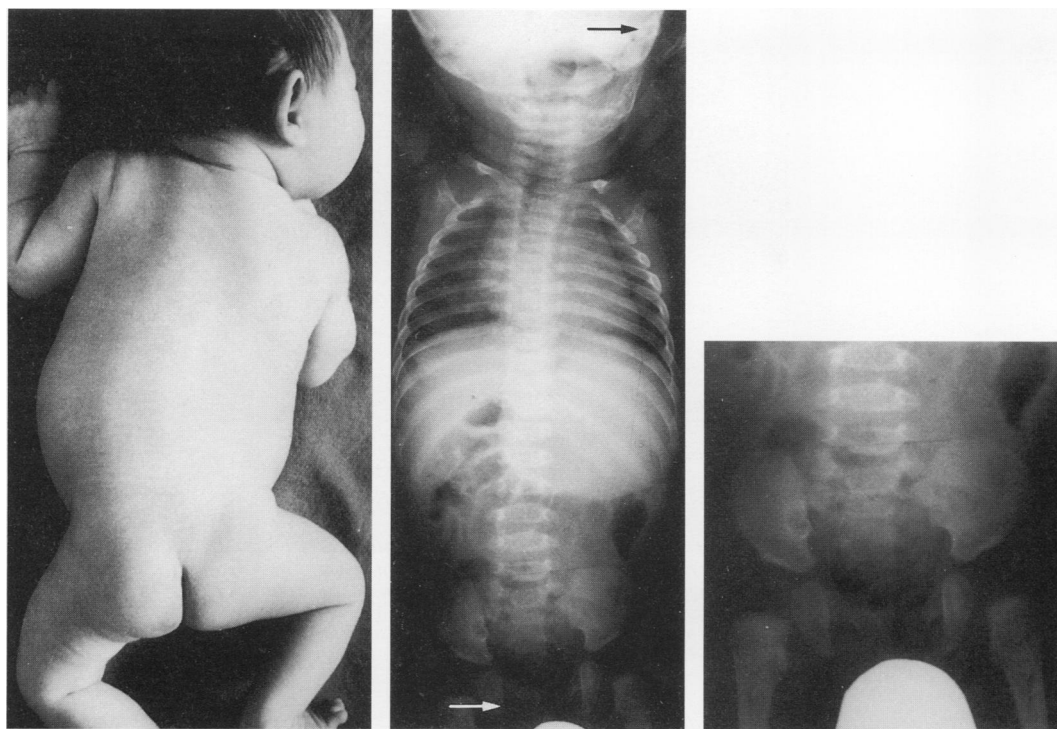
Figure 2 Sketch from a perspective inferior to the mother's spine of a child with TAC syndrome lying on the spine in left occipital position. The head is forced to the right and the left hip is adducted from the front axis line of the body.

deformities, consisting of four with torticollis, three with acetabular dysplasias, two with congenital dislocation of the hip, one with adduction contracture of the hip joint, one with scoliosis, and six with other asymmetrical postural deformities. The incidence of congenital dislocated hips in the siblings (5.3%) was greater than that of the general population in Japan (0.2–0.3%).<sup>13</sup> Only four among 124 parents had asymmetrical deformities: torticollis, congenital dislocation of the hip, acetabular dysplasia, and facial asymmetry. There are obvious difficulties in ascertaining whether these transient deformities were present in older relatives (grandparents, aunts, and uncles), and no instances of asymmetrical postural deformities were found in second degree relatives. Twelve (3.8%) of 318 first cousins had asymmetrical deformities (four with congenital dislocation of the hip, four with acetabular dysplasias, two with torticollis, one each with scoliosis and cleft lip), but the incidence was not significantly different from that for the general population.

Table 2 Relationship of the side of the torticollis and adducted hip contracture with fetal presentation

Presentation	Muscular torticollis		Adducted contracture	
	Left neck	Right neck	Left hip	Right hip
Left occipital*	7	0	41	10
Right occipital	0	7	6	29
	p<0.005		p<0.001	

\*Left occipital position: the back of the fetus is located on the left side of the mother and the left hip and left side of the neck of the fetus are situated on the mother's spine in vertex presentation.



**Figure 3** Occipital curve. Left: a view of the back showing thoracolumbar curvature convex to the occipital side. Centre: the black arrow indicates the facial side of the turned head. A spinal curvature convex to the occipital side is observed even when the skull is held in neutral rotation by the radiologist. Facial deformity, flatness on the facial side, is seen. The white arrow indicates the wider obturator foramen on the occipital side due to pelvic tilt. Right: a magnified view of pelvis indicating asymmetry of the obturator foramen.

#### CLINICAL COURSE FOR EACH SIGN IN THE TRIAD

The terms occipital side and facial side as used herein refer to the side of the patient's body corresponding to, respectively, the occipital and facial side of the head when in the rotated position.

##### 1. Tendency to turn the head to one side

Rotation of the head to the opposite side without any resistance could be performed in 85 patients, although it seemed to cause discomfort to the babies, who immediately reverted the head to the original position on release. In the other 23 patients, contracture and some resistance were felt when the head was passively turned to the opposite side. A palpable mass in the sternocleidomastoid muscle developed in 14 of these 23 patients within the first few weeks on the expected side, that is, the left if the fetus' left side rested on the mother's spine (table 2). All were successfully treated by single gentle full rotation of the head to the occipital side (manual myotomy) and by placing them in prone position when they were laid down. In one patient delivered by caesarean section, the rotated head position and contracture persisted for one month, although a palpable mass did not develop.

##### 2. (A) Adduction of the hip joints on the occipital side

All of the 108 patients showed adduction contracture of the hip on the occipital side of the turned head, which was defined as restriction of hip abduction in flexion compared with the angle of abduction on the facial side.

Radiographs usually demonstrated pelvic rotation toward the facial side, which was detected by increased transverse diameter of the obturator foramen on the occipital side (fig 3, right). In 81 patients, this limitation in abduction disappeared either spontaneously or by placing the patients in the prone position. Barlow's test was positive in two neonates and in five referred infants. In four neonates, Barlow's test was negative at the newborn examination but became positive within one to three months. In 16 patients unilateral acetabular dysplasia was demonstrated radiographically at 2 to 3 months of age. In one referred infant, the hip was stable with a negative Barlow's test at the first visit but was dislocated with positive Barlow's test and Ortolani's sign two months later. All of these 12 patients (two with congenital, five with suspected congenital, and five with late infantile dislocation) were successfully treated by placing the neonates in a prone position or by using the Pavlik harness for infants. At 4 years of age, four patients with previously dislocated hip showed unilateral acetabular dysplasia.

##### 2. (B) Abduction contracture of the hip joint on the facial side of the turned head

In 51 patients, the hip joint on the facial side was contracted in the abducted position, which was defined by tilting the gluteal fissure to the facial side in the prone position on bringing both legs together (fig 4, middle). Five required manual stretching of the contracture in order to correct persistent pelvic tilt and resulting lumbar scoliosis as described below.

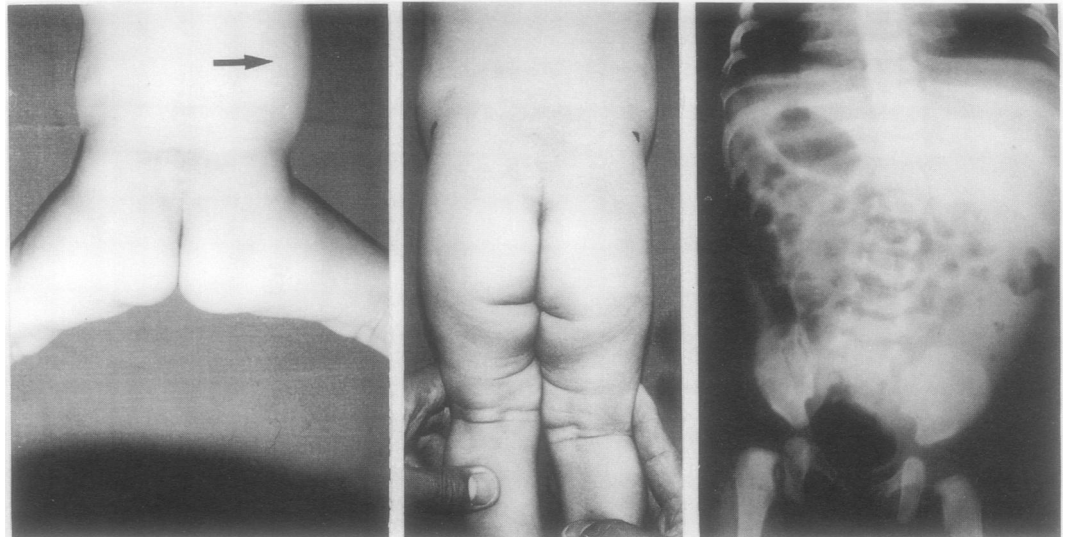


Figure 4 Abduction contracture of the hip joint observed in the patients with TAC syndrome. Left: the range of abduction of both hip joints. The arrow indicates the facial side of the turned head. Centre: the pelvis and gluteal fissure tilt to the facial side when the knees are brought together, giving the appearance of a leg length discrepancy. Right: the radiograph shows the spinal curvature convex to the facial side (facial curve) due to abduction contracture of the right hip with resulting pelvic tilt.

### 3. Truncal curvature

We observed three types of truncal curvature.

(A) All 54 neonatal cases showed a natural truncal curve which was convex in relation to the occipital side of the turned head. This type of curvature was also observed radiographically in 10 infantile cases (occipital curve) (fig 3, left, centre). The curve in most cases disappeared spontaneously within the first year, but in five (8%) it persisted for 18 months and in one for three years.

(B) In 18 infantile cases, abduction contracture of the hip joint on the facial side caused a pelvic tilt to the facial side when both legs were brought together, and lumbar postural scoliosis with the convexity also to the facial side ensued (facial curve) (fig 4, right). In four patients (22%) the curve was still present when the child started to walk, and in one it persisted until 3 years of age.

(C) A double curve (thoracic occipital and lumbar facial) was observed radiographically in 18 infants, in two of whom it was still present at 3 years of age.

### OTHER CLINICAL MANIFESTATIONS

We observed pes calcaneovalgus exclusively on the facial side in 12 babies; it was probably due to moulding by the uterine wall (fig 5). Forefoot adduction or supination exclusively on the occipital side was seen in four patients. Skull deformities were slight in the newborn period but were progressively aggravated if the head turning persisted and the patients were placed in the supine position. Facial asymmetry (straight on the facial side and round on the occipital side) was seen in 36 patients.

### Discussion

It has been reported that 15–20% of patients with torticollis also show dysplasia or dislocation of the hip joint,<sup>8,9</sup> and that 8–10% of those with congenital dislocation of the hip show torticollis,<sup>8,10</sup> mainly ipsilaterally. It has been speculated that the aetiology of this ipsilateral coexistence and also those with plagiocephaly<sup>11</sup> is intrauterine or partly postnatal. The subjects of these reports, however, were infants, and neither analyses of their neonatal condition or presentation of the fetus in utero nor comprehensive epidemiological surveys were carried out. Although congenital dislocation of the hip and torticollis are highly associated with breech presentation,<sup>3-5,14,15</sup> 65–90% of the babies with these conditions were delivered by a normal vertex presentation.<sup>9,12,16</sup> The aetiological factors and mechanisms of these deformations in patients delivered by a normal vertex presentation have not yet been fully explained. Dunn demonstrated that the leg resting on the mother's spine was more likely to be adducted and to be dislocated.<sup>4</sup> Our present epidemiological results suggest that TAC syndrome in a child delivered by vertex presentation is almost invariably associated with a tight uterus, and that both unilateral congenital dislocation of the hip and torticollis develop on the side of the fetus lying on the mother's spine (fig 2).

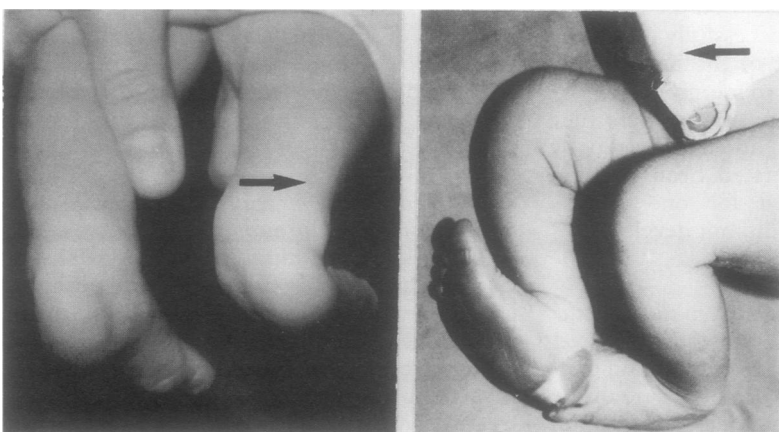


Figure 5 The arrows indicate the facial side of the turned head. Severe (left) and milder (right) pes calcaneovalgus of the right foot on the facial side is shown.

Conversely, the contralateral hip joint lying anteriorly showed the abduction contracture. We usually advised parents of babies with TAC syndrome to place them in the prone position, but some parents were reluctant to do so, and the asymmetrical deformities progressed. The five patients with late dislocations of the hip joint were among the latter, which suggests that persistent adducted contracture may inhibit the acetabular development, displace the femoral head laterally, and cause late infantile dislocation. The combination of adduction and abduction contractures of the hip joints in the same patient has been noted.<sup>17 18</sup> Postural scoliosis due to pelvic tilt caused by the adducted hip joints has been described by Tachdjian.<sup>19</sup> The clinical course of this type of curvature, however, has not been fully delineated. Although the natural ‘occipital’ curves in most patients resolved within a few weeks, as did the neck contracture, the ‘facial’ lumbar curves and the double curves associated with abduction contracture of the hip joint persisted longer, even until after the start of walking. A persistently turned fetal head towards the anterior wall of the uterus could well cause shortening,<sup>11</sup> disturbed venous circulation,<sup>20</sup> and contracture of the sternocleidomastoid muscle on the side of mother’s spine or on the fetal occipital side. We have also observed this contracture in a baby delivered by caesarean section. A palpable mass was found in 14 patients with TAC syndrome, eight of whom were among the 7103 examined newborns (incidence, 0.1%). A shortened and degenerated sternal branch in the sternocleidomastoid muscles would be vulnerable to stretching in the second internal rotation during vertex delivery, in which the head is rotated forcibly towards the maternal sacrum. Aggressive repair of these damaged muscles by granulomatous tissue would lead to the formation of a palpable mass. The associated findings of a high proportion of first born children, a greater proportion of births during the winter months, and higher maternal age suggest an increase in the tone of both the abdominal and uterine wall,<sup>3 5</sup> which would restrict movement of the fetus. The high incidence of several asymmetrical deformities in the siblings of index patients also suggests the influence of common intrauterine or intra-abdominal factors. Of the 7103 newborns, two developed congenital dislocation of the hip and four late infantile dislocation. Such late infantile dislocations would have been considered previously to be congenital dislocation which had been

overlooked. Of the 7103 newborns, apart from the patients with TAC syndrome, six had other congenital dislocation of the hip, which was mostly bilateral, and had a teratological, generally hypotonic or strong genetic background. The incidence of TAC syndrome in 7103 live births was 0.58%, and that of unilateral dislocation of the hip joint as a congenital postural deformity was 0.09% (two congenital plus four late dislocations in 7103), which represents half of the total cases of congenital dislocation of the hip in our neonates. This incidence is a third to half of that of congenital dislocation of the hip in Japan at present (0.2–0.3%). Improvement in maternal welfare and prenatal and postnatal education may lessen the incidence of TAC syndrome and resultant postural deformities still further.

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