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Congenital chest wall malformations in Nigerians

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Summary

The pattern of congenital chest wall malformations has been studied prospectively at the University of Ilorin Teaching Hospital between 1983 and 1987. A clinical series of 20 patients was found among 2195 patients selected randomly from 10,031 patients obtaining radiographs, and also from 1070 patients seen in the Cardiothoracic Surgery Clinic. An autopsy series of 15 foetuses and newborns was found among 205 studied for malformations. The 35 cases were found to have supernumerary ribs (10 clinical, eight autopsy), absent or defective ribs (three clinical, four autopsy), sternocostal deformities (three clinical, three autopsy) and four clinical cases with thoracic/thoraco–lumbar scoliosis and neonatal dwarfism with short ribs. Apart from two autopsy specimens (Cantrell's pentalogy with acrania and extensive abdominoperineal omphalocele) whose associated malformations appeared to be incompatible with life, all foetuses and newborns apparently had usual perinatal causes of death. One patient presented with costoclavicular syndrome while another had chest wall excision for a suspected tumour in a bifid rib. A patient with Poland's Syndrome and absent digits, and another infant with Cantrell's Syndrome are awaiting surgical reconstruction. The incidence of the malformations is approximately 1.01%. Other clinicopathological features are presented and the literature is reviewed.

Résumé

Une étude prospective (1983–87) a été menée sur la nature des malformations congénitales au centre hospitalo-universitaire d'Ilorin. Une série clinique de 20 malades a été tirée de 2195 malades repêchés au hasard parmi 10,031 personnes à la recherche des radiographies ainsi que de 1070 malades à la clinique de chirurgie cardiothoracique. Une série d'autopsie de 15 foetus et de nouveaux-nés a été découverte chez 205 sujets étudiés pour malformations. L'on a découvert chez les 35 sujets, des côtes supplémentaires (10 cliniques), huit d'autopsie) une absence ou une défectuosité de côtes (trois cliniques, quatre d'autopsie), des déformités sterno-costales (trois cliniques, trois d'autopsie) et quatre cas cliniques avec scoliose thoracique/thoracico-lombaire et nanisme néonatal avec côtes courtes. Exceptés deux échantillons d'autopsie (pentalogie de Cantrell avec acranie et omphalocèle abdominopérinéal étendu) dont les malformations accessoires semblaient incompatibles avec la vie, tous les foetus et nouveaux-nés manifestaient apparemment des causes périnatales habituelles de mort. Un malade a indiqué un syndrome costoclaviculaire alors qu'un autre avait subi une excision du mur de la poitrine pour cause soupçon de tumeur dans une côte bifurquée. Un malade souffrant du syndrome de Poland avec absence de doigts et un autre enfant atteint du syndrome de Cantrell attenent une reconstruction chirurgicale. L'incidence des malformations a été de 1.01% approximativement. D'autres traits clinico-pathologiques sont présentés alors que la littérature est passée en revue.

Introduction

The literature on congenital chest wall malformations in Africans is scanty. What is available is in the form of differential diagnosis in case reports [1], or mentioned with associated malformations [2–5]. On the other hand thoracic wall malformations were not described in five series that included extracardiac malformations [6–10].

This paper presents a series of 20 patients and 15 perinatal autopsies with varying degrees of

chest wall malformations. The paper is based on a study of 3265 patients and 205 perinatal autopsies done between 1983 and 1986 at the University of Ilorin and University of Ilorin Teaching Hospital (UITH).

Subjects and methods

A sample population of patients, attending the clinics and wards of the General Hospital Wing of the UITH between 1983 and 1986, was selected on the basis of having had at least one chest radiograph during the period of care. Out of this large sample patients with alternate odd-numbered X-ray records were selected for detailed review periodically during the study period. In addition, all patients attended to in the Cardiothoracic Surgery Clinic during the study period were similarly reviewed. Patients from these two sources presenting with chest wall malformations constituted the clinical series.

The autopsy series was collected from two sources between 1983 and 1986. After obtaining informed consent of parents, abortions, still-births and neonatal deaths were collected fresh-frozen from the Maternity Wing of the UITH and the ECWA Hospital, Egbe, about 100 km east of Ilorin. After documenting perinatal features and taking measurements of each specimen, each was studied for gross deformities. A longitudinal sternotomy incision was made and the viscera were removed for other studies. The ribs and vertebrae were then counted and all deformities were compiled. Overall incidence as well as incidence for each malformation was calculated.

Results

The total sample patient population was 10,031, with alternate odd-number selection of patients providing a theoretical sample of 2508 patients. Of these 2195 (87.5%) who had complete information were reviewed together with 1070 patients attending the Cardiothoracic Surgery Clinic to form a clinical series of 3470 patients. Twenty patients were found to have chest wall malformations. Twelve of these were male and eight female, producing a male:female ratio of 3:2. Ages ranged from new-born to 40 years (mean age ± s.d.: 15.6 ± 10.4).

One patient, an 18-year-old female, presented with symptoms suggestive of costoclavicular syndrome. Physical signs were, however, not convincing and there was no confirmatory angiography. A 2½-year-old female with a bifid right fifth rib (Figs 1 & 2) had resection of the rib and chest wall reconstruction because of clinical and radiological suspicion of tumour. No tumour was found during the operation or histologically. A 3-year-old boy with a bifid right third rib (Fig. 3) presented with swelling over the rib, he is being followed-up. A 2½-year-old boy with defective ribs presented with Poland's Synrome with associated right hand adactylia (Fig. 4). He has been followed from birth and is awaiting plastic surgery subject to clarification of cardiomegaly without murmur

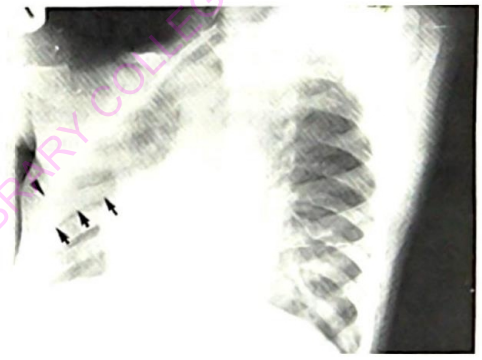

Fig. 1. Left anterior oblique chest radiograph of 2½-year-old girl with bifid rib and suspected tumour (†). (▲) point to posterior expanded portion of rib before bifurcation.

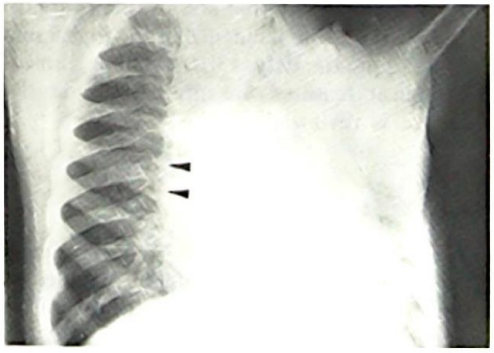

Fig. 2. Right anterior oblique chest radiograph of same patient as in Fig 1 showing anterior portion of expanded rib before bifurcation.

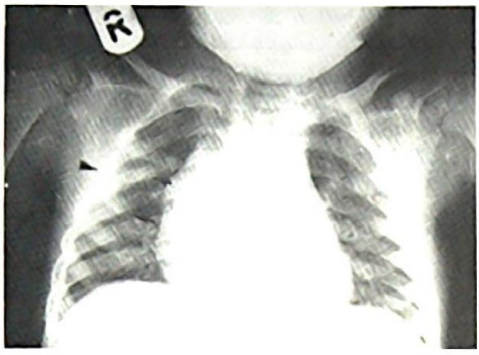

Fig. 3. Postero-anterior chest radiograph of 3-year-old boy with bifid anterior third rib (▲). Expanded rib before bifurcation (▲).

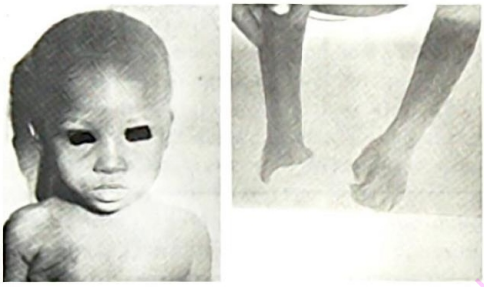

Fig. 4. Two-and-a-half-year-old boy with Poland's Syndrome (note the depressed right chest) and associated right adactylia.

being detected. A 5½-month-old female with Cantrell's pentalogy has a skin-covered omphalocele-like defect and dextrocardia with a ventricular septal defect murmur. She is thriving while awaiting definitive cardiac diagnosis.

Out of 205 foetuses and newborn corpses 15 were found to have chest wall anomalies. Ten of these had birth weights ranging from 1.2 to 2.5 kg with a mean of 1.9 ± 0.5 kg. While five others had weights ranging from 3.2 to 4.5 kg (mean 3.7 ± 0.5 kg). Three of the 15 were neonatal deaths, two were macerated stillbirths following intra-uterine death and compound presentation, six were fresh stillbirths due to obstetric causes and four were stillbirths with complex malformations. The latter included a set of thoraco-omphalopagus Siamese twins with a single heart and liver, a stillbirth with Cantrell's Syndrome, acrania and absent left upper limb (Fig. 5) and another with extensive

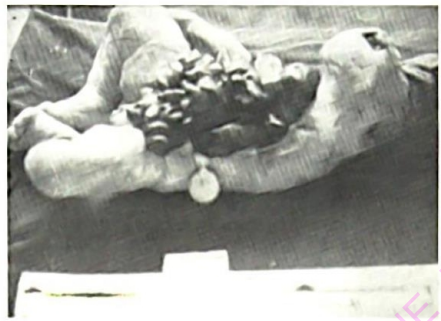

Fig. 5. Fresh stillbirth showing severe Cantrell's Syndrome with acrania and dysmelia of left upper limb.

sacroperineal omphalocele, absent left lower limb and malformed intestines associated with knobly 13th (lumbar) ribs. Apart from these unusual cases the foetuses and new-borns were very much like those not released for study as regards obstetric causes of death and external features.

The types of malformations and sex distribution for the entire series are shown in Table 1. The overall male:female ratio was 4:3 while overall incidence rate for malformations was 1.01%. The incident rate for each malformation is as follows:

(i) supernumerary ribs 0.52% (cervical ribs 0.20%, lumbar ribs 0.26%, bifid ribs 0.06%);

(ii) absent/defective/short ribs 0.20% (short ribs 0.14%, absent ribs 0.06%, rib defects 0.03%);

(iii) sternocostal deformities 0.17% (pectus carinatum 0.06%, Cantrell's Syndrome 0.06%, pectus excavatum was not found in this study); and

(iv) thoracic scoliosis 0.09%.

Discussion

The chest wall malformations presented in this series are well described in standard textbooks. However, there are certain peculiarities worth mentioning. The paper presents a series as well as incidence rates in black Africans. As the majority of patients were selected randomly the figures are probably reliable. Pectus excavatum which is regarded as the most common sternal deformity [11] is conspicuously absent, but the

Table 1. Chest wall malformations — types and distribution according to sex

Malformations	Clinical			Autopsy			Total
	Male	Female	Total	Male	Female	Total	
Supernumerary ribs	6	4	10	6	2	8	18
Cervical	4	3	7				7
Bifid	1	1	2				2
Lumbar	1		1	6	2	8	9
Absent/defective ribs	3		3	2	2	4	7
Short				2	2	4	4
Absent	2		2				2
Defective	1		1				1
Sternal and sternocostal	1	2	3		3	3	6
Thor-omph*					2	2	2
Pectus carinatum	1	1	2				2
Cantrell's Syndrome		1	1		1	1	2
Others	2	2	4				4
Thoracic scoliosis	2		2				
T/L scoliosis		1	1				
Neonatal dwarfism		1	1				
Total	12	8	20	8	7	15	35

*Thoraco-omphalopagus Siamese twins.
†Thoraco-lumbar.

reason for this is not clear. It may be due to the fact that it presents at a relatively younger age than pectus carinatum [11]. It may therefore not be striking enough for parents to notice the less severe forms as they grow imperceptibly with the young child. As it is hidden under the clothes, the older child with pectus excavatum may easily compensate by avoiding exposure of the deformity. Also pectus excavatum is frequently and erroneously thought to produce no physiological derangements. However, pulmonary function tests carried out in the erect position and during excercise [11–13], and cardiac catheterization done in the sitting rather than supine position and during exercise revealed significant derangements [11]. It is understandable that in an environment where many are pre-occupied with basic health problems few people pay attention to deformities that are not severely or moderately incapacitating at rest.

Our 0.20% incidence of cervical ribs compares favourably with 0.25% of routine thoracic roentgenograms found by Ravitch [11] although lower than that reported by Cockshott in Ibadan [14]. It has been estimated that, although found in about 0.5% of the general population, only about 10% of such people are symptomatic [15]. One patient in our series, with symptoms suggestive of costoclavicular syndrome, presented with neither convincing physical signs nor clear indications for angiography. A larger patient population would be expected to contain symptomatic patients requiring diagnostic and therapeutic procedures.

The absence of cervical ribs in our autopsy series appears to have an embryological basis as described by Glenn *et al.* [12] and Hamilton and Mossman [16]. The sclerotome differentiates from somites to form the axial skeleton while the ribs develop following chondrification in the adjacent membranous costal area. At the level of the 7th cervical vertebra an ossific centre which appears in the 6th intra-uterine month forms the anterior portion of the transverse process of the 7th cervical vertebra which in turn fuses with the posterior portion in the 5th or 6th year. It would, therefore, appear that the

short and soft cartilaginous would-be cervical ribs might be missed at autopsy. Extrathoracic musculo-skeletal malformations not infrequently found in association with chest wall malformations appear to represent an extension of the same maldevelopment.

Although cervical ribs produce symptoms in a small percentage of cases, lumbar ribs and absent or deficient lower thoracic ribs may become clinically significant in victims of thoraco-abdominal trauma. However, Rickham [17] has reported lung hernia secondary to congential absence of ribs.

Associated malformations affect management and prognosis of chest wall malformations significantly, for example the various anomalies associated with Cantrell's pentalogy [11,18–20]. The more severe malformations are incompatible with life as was observed in three or four of the autopsy specimens presented here. Theoretically one of the Siamese twins could have been saved operatively. However, they were delivered stillborn by Caesarian section, the indication for which was compound presentation with four limbs.

There was no known aetiology for the malformations. There was no known thalidomide-like tranquilizers ingested during pregnancy, but the mother of the stillbirth presenting with severe Cantrell's Syndrome admitted to using a traditional bathing concoction during pregnancy. Many more women probably ingest traditional drugs in pregnancy. Some of the musculo-skeletal abnormalities seen in the autopsy series are not too different from those presented by the thalidomide babies [21].

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References

1. Grillo IA, Johnson A, Akinyemi OO, Ladipo OA. Congenital diaphragmatic hernia simulating pectus carinatum in a two-year old boy. Nig Med J 1977;7:210–15.
2. Antia AU. Familial skeletal cardiovascular syndrome (Holt Oram) in a polygamous African family. Br Heart J 1970;32:241–4.
3. Adeyokunnu AA, Akingbehin NA. Experience with the management of exomphalos and gastroschisis in Ibadan. Nig J Paediatr 1981;8:45–51.
4. Adekunle OO, Johnson AOK. Congenital anorectal anomalies in Western Nigeria. Nig J Paediatr 1981;8:40–4.
5. Anyanwu CH, Okoroma EO, Ihenacho, HNC, Umeh BU. Experience with surgical management of cardiovascular diseases in children. Nig J Paediatr 1981;8:94–7.
6. Gupta B, Antia AU. Incidence of congenital heart disease in Nigerian children. Br Heart J. 1967;29:906–9.
7. Antia AU, Williams AO. Congenital heart disease in Nigeria. Necropsy study of 47 cases. Br Heart J. 1971;33:133–7.
8. Holmes GE. Observations on 1500 consecutive African births in East Africa. E Afr Med J 1973; 50:498–513.
9. Okeahealam TC. The pattern of congenital malformations observed in Dar es Salaam. E Afr Med J 1974;51:101–8.
10. Jaiyesimi F, Antia AU. Extracardiac defects in children with congenital heart disease Br Heart J 1979;42:475–9.
11. Ravitch MM. Disorders of the sternum and the thoracic wall. In: Sabiston DC. Jr, Spencer FC, eds. Surgery of the Chest. Philadelphia: WB Saunders, 1976:324–69.
12. Glenn WWL, Liebow AA, Lindskog GE. Diseases of the chest wall. In: Thoracic and Cardiovascular Surgery with Related Pathology. New York: Appleton-Century-Crofts, 1975: 117–140.
13. Weg JG, Krumholz RA, Harkleroad LE. Pulmonary dysfunction in pectus excavatum. Am Rev Respir Dis 1967;96:936–45.
14. Cockshott WP. Anatomical anomalies observed in radiographs of Nigerians. W Afr Med J 1958; 7:179–84.
15. Imparato AM, Ryles TS. Peripheral arterial disease. In: Schwartz SI, ed. Principles of Surgery. New York: McGraw-Hill, 1984:897–974.
16. Hamilton WJ, Mossman HW. Skeletal system. In: Hamilton, Boyd and Mossman's, Human Embryology. Williams and Wilkins, 1972:526–47.
17. Rickham PP. Lung hernia secondary to congeni-

tal absence of ribs. Arch Dis Child 1959;34:14–17.
18. Cantrell JR, Haller JA. Jr, Ravitch MM. A Syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart. Surg Gynecol Obstet 1958; 107:602–14.
19. Mulder DG, Crittenden IH, Adams FH. Complete repair of a syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart; excision of left ventricular diverticulum. Ann Surg 1960;151; 113–22.
20. Gula G, Yacoub M. Syndrome of congenital ventricular diverticulum and midline thoracoabdominal defects. Thorax 1977;32:365–9.
21. Hamilton WJ, Mossman HW. Determination, differentiation, the organizer mechnism, abnormal development and twinning. In: Hamilton, Boyd and Mossman's, Human Embryology. William and Wilkins, 1972:192–227.

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