UNIVERSITI PUTRA MALAYSIA

CONGENITAL KYPHOSIS AND KYPHOSCOLIOSIS: THE NATURAL HISTORY AND RESULTS AFTER SURGERY

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CONGENITAL KYPHOSIS AND KYPHOSCOLIOSIS : THE NATURAL HISTORY AND RESULTS AFTER SURGERY

By

HARWANT SINGH

Thesis Submitted in Fulfilment of the Requirements for the Degree of Doctor of Philosophy in The Faculty of Medicine and Health Sciences, Universiti Putra Malaysia

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DEDICATION

This thesis is dedicated to my parents, to whom I owe everything; and to my wife, who put up with my nuances while I was doing this study.

It is also dedicated to the late Professor N. Subramaniam of the University of Malaya, and the late Professor Q.M. Iqbal of Universiti Kebangsaan Malaysia; who were the pioneers of scoliosis surgery in Malaysia, and who taught me that treating children's spinal deformities is a life long passion and not just a vocation.

Lastly, but by no means least; this is also dedicated to all the spinal deformity patients who have given me the opportunity to treat them, and who have taught me so much more than any book, conference or meeting; and have given me valuable insight into what spinal deformity is.

Abstract of thesis presented to the Senate of Universiti Putra Malaysia in fulfilment of the requirements for the degree of Doctor of Philosophy.

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October 2000

Chairman: **Professor Dr Leslie Lai Chin Loy**

Faculty: **Medicine and Health Sciences**

Congenital kyphosis and kyphoscoliosis are due to vertebral anomalies that are present at birth. These vertebral anomalies cause a deformity of the spine in the sagittal plane. If allowed to progress, this deformity can produce neural dysfunction due to bowstringing of the cord over the apex of the deformity. The goals of this study were to document the natural history of congenital kyphosis and kyphoscoliosis and to determine the stage at which the natural history should be interrupted by treatment. This was done by reviewing the medical records and radiographs of the spine of 112 consecutive patients. Sixty eight patients had a type-I kyphosis due to anterior failure of vertebral-body formation, twenty-four had a type-II kyphosis due to anterior failure of vertebral-body segmentation, and twelve had a type-III kyphosis due to a combination of anomalies. The deformities of the remaining eight patients could not be classified. The apex of kyphosis was seen at all

levels but was most frequent between the tenth thoracic and the first lumbar level (seventy-four patients; 66%). Progression of the curve was most rapid during the adolescent growth spurt and only stopped at skeletal maturity. Progression was most rapid and the magnitude of the curve was the greatest in the type-III kyphosis (twelve patients) followed by type-I kyphosis due to posterolateral quadrant vertebra (thirty-nine patients), a posterior hemivertebra (eight patients), a butterfly vertebra (fifteen patients), and a wedge vertebra (six patients). A kyphosis due to two adjacent type-I vertebral anomalies progressed more rapidly and produced a more severe deformity than a single similar anomaly. The prognosis for type-II kyphosis was variable and was much more severe when an anterolateral unsegmented bar had produced a kyphoscoliosis (nine patients) than when a midline anterior bar had produced a pure kyphosis (fifteen patients), which usually progressed slowly. Sixty-five patients had surgical treatment. The stability of the sagittal curve at maturity was dependent on vertebral anomaly causing sagittal deformity, type of arthrodesis procedure performed, age of patient at arthrodesis and size of sagittal curve at arthrodesis, however only the first two were statistically significant.

Abstrak tesis yang dikemukakan kepada Senat Universiti Putra Malaysia sebagai memenuhi keperluan untuk ijazah Doktor Falsafah

KYPHOSIS DAN KYPHOSCOLIOSIS KONGENITAL: RIWAYAT SEMULAJADI DAN KESIMPULAN SURGERI

Oleh

HARWANT SINGH

Oktober 2000

Pengerusi: **Professor Dr Leslie Lai Chin Loy**

Fakulti: **Perubatan dan Sains Kesihatan**

Kyphosis dan kyphoscoliosis kongenital adalah disebabkan oleh malformasi vertebra yang wujud dari lahir. Malformasi ini menyebabkan deformiti spina di plana sagital. Jika deformiti ini berterusan, saraf tunjang boleh dikepil oleh apex deformiti menyebabkan kehilangan fungi neural. Tujuan kajian ini adalah untuk meneliti kejadian deformiti ini, dan mengesyorkan rumusan untuk rawatan penyakit ini. Kajian telah dijalankan dengan penelitian nota-nota perubatan dan radiograf-radiograf spina 112 pesakit konsekutif. Enam puluh lapan pesakit mengalami kyphosis Jenis 1, dua puluh empat mengalami kyphosis Jenis 2, dan dua belas mengalami kyphosis Jenis 3. Baki lapan pesakit tidak dapat diklassifikasikan.

Apeks kyphosis di lihat di semua tahap, tetapi didapati sering diantara vertebra thorasik kesepuluh dan vertebra lumbar pertama (tujuh puluh empat peaskit; 66%). Kepecutan deformiti ini adalah terlaju semasa waktu baligh dan hanya berhenti selepas kebalighan rangka tercapai. Kepecutan deformiti ini adalah terlaju dan magnitud deformiti adalah terbesar di kyphosis Jenis 3. Kyphosis yang disebabkan oleh dua anomali vertebra bersampingan mempunyai kepecutan deformiti yang lebih laju; dan menyebabkan deformiti yang lebih besar magnitud berbanding dengan anomali satu vertebra sahaja. Prognosis kyphosis Jenis 2 adalah lebih baik dengan kepecutan deformiti yang kurang dari Jenis 1 atau 3.

Enam puluh lima pesakit telah menjalankan rawatan pembedahan. Stabiliti deformiti pada waktu baligh adalah bergantung kepada jenis anomali vertebra yang menyebabkan deformiti sagital, jenis artrodesis yang dijalankan, umur pesakit pada waktu pembedahan dijalankan dan saiz deformiti pada masa pembedahan; tetapi hanya faktor pertama dan kedua adalah signifikan.

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The President and Fellows of The Royal College of Surgeons of Edinburgh, for permission to reproduce the illustrations in Figures 1, 2, 4, 5, 6, 7, 8 and 9.

I certify that an Examination Committee met on 30 October 2000 to conduct the final examination of Harwant Singh on his Doctor of Philosophy thesis entitled "Congenital Kyphosis and Kyphoscoliosis: The Natural History And Results After Surgery" in accordance with Universiti Pertanian Malaysia (Higher Degree) Act 1980 and Universiti Pertanian Malaysia (Higher Degree) Regulation 1981. The Committee recommends that the candidate be awarded the relevant degree. Members of the Examination Committee are as follows:

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This thesis submitted to the Senate of Universiti Putra Malaysia has been accepted as fulfilment of the requirement for the degree of Doctor of Philosophy.

KAMIS AWANG, Ph.D.
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Date: 11 JAN 2001

DECLARATION

I hereby declare that the thesis is based on my original work except for quotations and citations which have been duly acknowledged. I also declare that it has not been previously or concurrently submitted for any other degree at UPM or other institutions.

Harwant Singh,

Date: 12 November 2000

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GLOSSARY

ABBREVIATIONS

MRI – Magnetic Resonance Imaging

FVC – Forced Vital Capacity

FEV1 – Forced Expiration in 1 second

CT – Computerized Axial Tomogram

BIOMECHANICS

Bending – Angular deformation of a structure, caused by a bending moment.

Bending Moment – The moment that tends to bend a structure. It is usually the sum of the moments due to several forces.

Centre of Gravity – The point in a body in which the body mass is centred.

Compression Force – A force that tends to shorten a structure or material.

Compressive Stress – A normal stress that tends to shorten material.

Force – An action that causes a body to displace or deform (Newtons – N).

Moment – The sum of the forces applied to a structure multiplied by their perpendicular distance from a reference point or axis (Newton meters – Nm).

Tension Force – A force that tends to elongate a structure or material.

Tensile Stress – A normal stress that tends to elongate material.

Viscoelasticity – Material behaviour in which the resistance to deformation depends on the amount of deformation (elastic) and the rate of deformation (viscous).

CLINICAL

Apical disc – The disc most deviated from the vertical axis of the patient.

Apical vertebra – The vertebra most deviated in the vertical axis of the patient.

Congenital scoliosis – Scoliosis due to congenitally anomalous vertebral development.

End vertebra – The most cephalad vertebra of a curve, whose superior surface or transverse axis, and the most caudal vertebra, whose inferior surface of transverse axis, tilts maximally towards the concavity of the curve.

Iliac apophysis – The apophysis along the crest of the ilium.

Kyphosis – A posterior convex angulation of the spine.

Kyphoscoliosis – A non idiopathic scoliosis associated with an area of hyperkyphosis.

Neuromuscular scoliosis – A scoliosis due to either a muscular or neurologic disorder.

Risser sign – In the frontal plane of the pelvis, the state of ossification of the iliac apopysis is used to denote the degree of skeletal maturity.

Sagittal balance – Alignment of C7 to the posterior superior aspect of the sacrum on an upright long cassette radiograph of the spine.

Scoliosis – A lateral curvature of the spine.

Skeletal age – The age obtained by comparing PA radiographs of the left wrist and hand with standards of the Gruelich and Pyle atlas.

Vertebral tilt – Angulation in the coronal plane, measured from the lower endplate to the horizontal.

CHAPTER 1

INTRODUCTION

1.1 General

Congenital Kyphosis, by definition, is an anatomical vertebral anomaly which is present at birth. This anatomical vertebral anomaly forms in utero and may manifest at birth, or soon after birth. There may be other congenital abnormalities associated with it. The early occurrence of the curve results in a potentially progressive curve, as the child will have progression during the physiological growth spurts.

These curves are usually rigid, resistant to correction by external bracing; and require surgery usually to arrest their progression. If the early curves are missed at birth or in early life, severe curves develop (Figure 1). It is more difficult to treat a severe rigid curve, than a minor curve. Therefore a proper and thorough understanding of the natural history of these curves is essential, to decide the appropriate time of treatment.

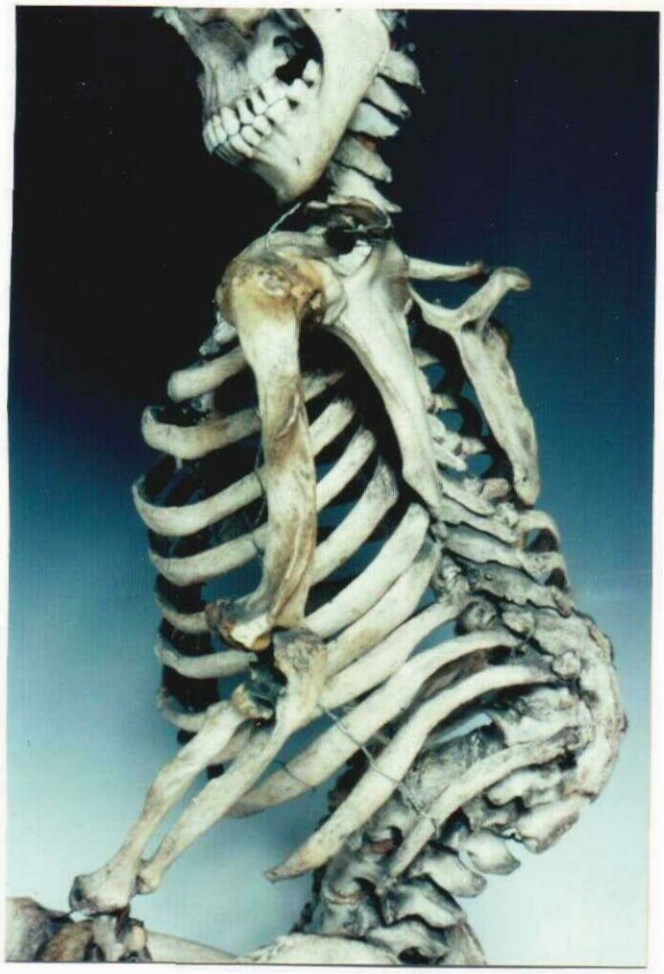

Figure 1: Kyphosis at the thoraco-lumbar level.

1.2 Historical Perspective

Spinal problems have been recognised in written history as far as 4500 years ago (Hughes 1988). Kyphosis, as a distinct pathological condition of the spine had been described by Hippocrates (Adams 1849, 1888), more than 2000 years ago; and surgical intervention as treatment for spinal disorders had been prescribed in early Indian surgical texts (Cumston 1926) approximately 1500 years ago. Although the clinical condition of kyphosis was recognised since early times, these descriptions could not determine the cause of the deformities. The anatomical works *De Dissectione Partium Corporis Humani* of Charles Estienne in 1539 and *De Humani Corporis Fabrica Libri Jeptum* by Andreas Versalius in 1543 contain the first modern and accurate anatomical descriptions of the human spine (Ball 1928). These works were the beginnings of serious study. However, there was no distinction between infective and non-infective causes of the kyphosis deformity of the spine.

The first description of congenital kyphosis as a separate entity causing spinal kyphosis was in German by Von Rokitansky in 1844. MacEwen first reported the association of untreated spinal deformity and neurological deficit in 1888. The development of radiography in 1895 enabled spinal deformities to be