

THE DEVELOPMENT OF IDEAS ON THE CARDIAC AND RESPIRATORY COMPLICATIONS OF SPINAL DEFORMITIES

by

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INTRODUCTION

THE CARDIAC and respiratory difficulties of patients with spinal deformities are readily apparent to the physicians who deal with them and have attracted a host of interpretations over the last two millennia. This is, no doubt, in part due to the prominence of the deformity, which renders the sufferer easily distinguishable from the normal. In the past, too, the prevalence of conditions such as Pott's disease of the spine was greater than today, and correspondingly more attention was focused on them.

Initially attention was concentrated on the symptomatic disability of these subjects, especially their shortness of breath. Later, the common occurrence of right ventricular hypertrophy and the small size of the lungs were emphasized. Physiological investigations have confirmed the latter and demonstrated the pulmonary hypertension which results from it.

EARLY VIEWS

Hippocrates (c. 460–375 B.C.) was aware of the cardio-respiratory problems of the hunchback. In his forty-sixth aphorism he writes: "Such persons as become humpbacked from asthma or cough before puberty, die",¹ and in paragraph 41 of his *On articulations* he again emphasizes that most patients with a gibbosity "above the diaphragm" die before sixty. He comments that "the cavities which inspire and expire the breath do not attain their proper capacity" and that the sufferers "become affected with difficulty of breathing and hoarseness".²

In the seventeenth century Severinus³ in Frankfurt and Floyer⁴ in England both mentioned the respiratory symptoms of the gibbous. At this time both the terms "gibbus" and "asthma" were used in slightly different senses from today. Although some authors confined the use of "gibbus" to an antero-posterior spinal deformity, usually kyphosis, others included scoliosis. The term "asthma" implied laborious

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¹ *The genuine works of Hippocrates, Translated from the Greek with a preliminary discourse and annotations by F. Adams*, London, Sydenham Society, 1849, vol. 2, p. 760.

² *Ibid.*, pp. 604–605.

³ M. A. Severinus, *De recondita abscessuum natura*, Libri 6: *De gibbis, valgis, varis, et aliis ab interna vi varie luxatis*, Frankfurt, J. Beyer, 1643, pp. 320–366.

⁴ J. Floyer, *A treatise of the asthma*, London, R. Wilkin, 1698, p. xxvii.

and difficult breathing, often noisy and wheezy, but with no specific connotation of bronchial obstruction.

Severinus⁵ discussed Hippocrates' sayings at length and altered the emphasis of his aphorism. He felt that the gibbosity was not necessarily caused by asthma, or cough, but that if gibbosity, cough, and asthma all occurred together, death was likely to occur before puberty. Floyer in his *Treatise of the asthma* recognized gibbosity as a cause of "continued asthma",⁶ and continued: "the gibbous are asthmatic, because of the contortion of the spinal marrow, the compression of the nerves, or the ill shape of the cavity of the breast, which straitens the lungs."⁷

In the eighteenth century Haller, too, was puzzled by Hippocrates' idea that a "dislocation of the vertebrae" could result from asthma and cough.⁸ He agreed with Severinus but added that the symptoms could also be due to the deformity, which may be so slight as to be unnoticed. However, if it was severe enough to cause asthma and cough in infancy, life past puberty was rare because of the damage to the heart and lungs. In defence of Hippocrates, though, he mentioned that *pectus excavatum* could result from the stretching and twisting of the bones during intense respiratory efforts and asthma.

Sauvages⁹ described "asthma a gibbo" in his nosology but agreed with both Hippocrates and Haller in saying that "not only does the gibbosity arise from asthma, and if this is so, death before puberty is likely, but from the gibbosity many suffer from asthma."

However, a generation before these deliberations, Petit had made considerable advances in a treatise on rickets written in France in 1723 and soon translated into English.¹⁰ He attributed the difficulty in breathing first to the "distortion of the spine [which] alters the disposition of the ribs and the direction of the muscles that influence them", and second to the smallness of the lungs which was caused by the sinking-in of the ribs and by the liver and spleen rendering the diaphragm convex.

He wrote that the force of the heart was increased and that "the blood is sent from the heart to the lungs with more ease than it is return'd from the lungs to the heart, which is no small cause of the disorders that happen therein". He felt that the "heaviness and grossness [of the blood] will be apt to cause some obstruction in the capillary vessels of the substance of the lungs". This "ill-quality of the blood", he postulated, was due to inspiration being hindered and would cause it to "stagnate in the capillary vessels of the pulmonick veins and arteries, upon finding the least disposition thereunto in these organs."

Unfortunately this speculative account was completely ignored by later writers.

⁵ Severinus, *op. cit.*, note 3 above.

⁶ Floyer, *op. cit.*, note 4 above, p. xxvii.

⁷ *Ibid.*, p. 131.

⁸ A. von Haller, *Disputationes ad morborum historiam et curationem facientes*, vol. 2, part 2: *Morbi pectoris*, Lausanne, Marci-Michael, 1757, p. 138.

⁹ F. B. de Sauvages, *Nosologia methodica, sistens morborum classes, genera et species, juxta Sydenhami mentem et botanicorum ordinem*, part 2, vol. 2, Amsterdam, Fratrum de Tourne, 1763, p. 177.

¹⁰ J.-L. Petit, *A treatise of the diseases of the bones; containing an exact and compleat account of the nature, signs, causes, and cures thereof, in all their various kinds*, London, T. Woodward, 1726, pp. 480-482.

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Several did, however, make interesting observations. Levacher de la Feutrie¹¹ described two girls with severely curved spines, deformed chests, and shallow respirations. Cullen, in his extensive nosology, gave “asthma a gibbo” as a cause of dyspnoea under the heading “ex compresso pulmone a partibus thoracem cingentibus”.¹² In a later work, he describes it as “dyspnoea thoracica” and states that there is no cure, but that palliation may “be obtained chiefly by avoiding a plethoric state of the lungs, and every circumstance that may hurry respiration”.¹³

On the other hand, Ludwig, writing from Leipzig, recollected one of his fellow soldiers with a severe gibbosity who frequently raced against other soldiers, and despite panting hard, mocked Ludwig’s warnings of the dangers of haemoptysis or some other disease supervening.¹⁴ Perhaps as a consequence of this experience, Ludwig recommended that “if at all possible exercise should be taken in the open air as this benefits the replenishment of the blood in the lungs”.¹⁵

All these authors had been concerned only with observing the symptoms and natural history of the patient with a spinal curvature. However, Morgagni, in his famous work *The seats and causes of diseases*, correlated his clinical observations with detailed autopsy findings. He recorded several patients with spinal deformities. Among these was a man aged twenty-nine, with a scoliosis convex to the left, whose autopsy showed a very narrow thoracic cavity on the left, a rather large heart, and a “kind of foam in the bronchia”.¹⁶ Similarly, a seventy-one-year-old cardinal had the cavity of one-half of his chest much smaller than the other, with a correspondingly smaller lung.¹⁷ A fifty-year-old beggar, who had a scoliosis convex to the left, died of an intracerebral haemorrhage. Morgagni postulated that this was due to inflection of the aorta causing excessive blood to be redirected to the upper part of the body.¹⁸

At this time interest in spinal curvatures was awakening in England. Percivall Pott, in a monograph on paraplegia following spinal deformity published in 1782, commented that a “hard dry cough, laborious respiration, quick pulse, and disposition to hectic, appear pretty early, and in such a manner as to demand attention”.¹⁹ A year later, Sheldrake emphasized that “if the thorax is much distorted, the lungs will be compressed” and that “the constitution will be gradually destroyed in a few years, if a consumption is not immediately produced by it [the deformity]”.²⁰ In the second

¹¹ Levacher de la Feutrie, *Traité du raktis ou l'art de redresser les enfants contrefaits*, Paris, Lacombe, 1772, pp. 426–427.

¹² W. Cullen, *Synopsis nosologiae methodicae in usum studiosorum*, 2nd ed., Edinburgh, A. Kincaid & W. Creech, 1772, p. 338.

¹³ W. Cullen, *First lines of the practice of physic, for the use of students in the University of Edinburgh*, 4th ed., Edinburgh, C. Elliott, 1784, vol. 3, p. 384.

¹⁴ C. G. Ludwig, *Adversaria medico practica*, Leipzig, Weidmann & Reich, 1771, vol. 2, part 4, p. 618.

¹⁵ *Ibid.*, p. 592.

¹⁶ G. B. Morgagni, *The seats and causes of diseases, investigated by anatomy*, translated by B. Alexander, London, A. Millar, 1769, vol. 1, pp. 73–74.

¹⁷ *Ibid.*, pp. 275–277.

¹⁸ *Ibid.*, vol. 3, p. 478.

¹⁹ P. Pott, *Further remarks on the useless state of the lower limbs, in consequence of a curvature of the spine: being a supplement to a former treatise on that subject*, London, J. Johnson, 1782.

²⁰ T. Sheldrake jr., *An essay on the various causes and effects of the distorted spine*, London, Dilly, Lewis & Faulder, 1783, p. 10.

volume of his encyclopaedic work, *Zoonomia*, Erasmus Darwin quoted a ten-year-old girl whose rapid respirations and palpitations improved as the spinal distortion was corrected with rest and “the bark”.²¹

RESPIRATION

Jones, in 1788, was the first person since Petit (1726)²² to advance any theories explaining the origin of the respiratory difficulties. He recorded the small size of the lungs and, describing a case, he wrote: “The ends of the ribs on the left side are strangely crowded on one another. This will produce inaptitude for motion; in such cases consequently the patient is fatigued, and out of breath with very little exercise”.²³

The concept that the lungs were small and expanded poorly because of the deformed thoracic cage was extended by Bampfield.²⁴ He analysed carefully the distortion of the thoracic skeleton and concluded that the abnormal position and lengths of the muscles decreased their power to dilate the cavity of the chest. He concluded that the consequent short laborious respirations led to “dyspnoea, asthma, congestion of the lungs, and defective oxygenation of the blood”. He found that the lungs of two patients who died during attacks of asthma were “gorged with black blood”.²⁵

Delpech also commented that the lungs were progressively “restrained” by the bones of the chest, which were so fixed that the muscles could hardly move them. He also described a patient whose breathlessness was, he proposed, the result of the stretching of the nerves to the diaphragm by the deformity.²⁶ Forget²⁷ emphasized that the diaphragmatic movement could be limited by the liver, which was frequently enlarged due to stasis of blood. He postulated, too, that the continual exaggerated inspiratory efforts led to pulmonary emphysema.

Guérin, in a prize-winning treatise submitted to the Académie des Sciences in Paris reported by Double,²⁸ described how the lungs “passed successively through the stages of engorgement, splenization, and carnification, and are even partially transformed into a fibro-cellular tissue”. These changes, he postulated, led to incomplete “arterialization” of the blood which was insufficient for the nutrition of the body. In contrast, Bouvier maintained that the mechanical action of respiration was imperfect because of “changes in the direction, form, and mobility of the ribs and

²¹E. Darwin, *Zoonomia; or the laws of organic life*, London, J. Johnson, 1796, vol. 2, pp. 87–92.

²²Petit, *op. cit.*, note 10 above.

²³P. Jones, *An essay on crookedness, or distortions of the spine; showing the insufficiency of a variety of modes made use of for relief in these cases; and proposing methods, easy, safe, and more effectual for the completion of their cures; with some hints for the prevention of these affections, and their disagreeable, painful, and dangerous consequences*, London, S. Gosnell, 1788, p. 140.

²⁴R. W. Bampfield, *An essay on curvatures and diseases of the spine, including all the forms of spinal distortion*, London, Longman, Hurst, Rees, Orme, Brown & Green, 1824, pp. 20–21.

²⁵*Ibid.*, pp. 48–50.

²⁶J. Delpech, *De l'orthomorphie par rapport à l'espèce humaine ou recherches anatomico-pathologiques sur les causes, les moyens de prévenir, ceux de guérir les principales difformités et sur les véritables fondemens de l'art appelé: orthopedique*, vol. 1, Montpellier, J. Martel, Jr., 1828, pp. 348–366.

²⁷C. P. Forget, ‘Considerations sur certaines asphyxies par cause organique’, *J. hebd. Prog. Sci. méd.*, 1836, 4: 161–176.

²⁸F. J. Double, ‘Rapports de la commission pour le grand prix de chirurgie’, *C. r. Séanc. Acad. Sci., Paris*, 1837, 5: 230–259.

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by difficulty in lowering the diaphragm”,²⁹ a view similar to that of Bampfield.³⁰ Bouvier also commented on the incomplete “arterialization” of the blood and on the rapid respirations that occurred on the slightest exertion.

HEART AND CIRCULATION

In the eighteenth century theories regarding the functioning of the heart in spinal deformities remained primitive. Jones felt, like Morgagni,³¹ that the curved aorta obstructed flow to the lower half of the body, and that “the force of the heart [was] weak for want of room”.³² Donner, in his M.D. thesis at Göttingen, argued also that the heart was “twisted in various ways, displaced, and impeded in [its] functions”,³³ and Vrolik attributed the curvature of the aorta to the displacement of the heart rather than to the aorta following the curvature of the spine.³⁴ Guérin, in his prize-winning treatise, also mentioned the mechanical effects of spinal deformity on the heart.³⁵ He felt that if the spinal deviation were to the right the great vessels were distorted at their origin, and when to the left, the movements of the heart “may become completely impossible”.

Corvisart, the famous French clinician, published an account of a thirty-six-year-old man whose aorta closely followed the course of the severely curved spine.³⁶ He too concluded that the aortic obstruction led to the enlargement of the right heart which he had observed. He argued that the interpolation of a capillary system between the obstruction and the hypertrophy was quite possible, but he failed to explain why the left side of the heart was normal.

This idea of obstruction of the large vessels causing cardiac hypertrophy was carried a stage further by Harrison in 1820. He felt that the pulmonary arteries as well as the aorta could be so distorted as to throw a strain on the heart “which, from a curious provision of nature, increases in size and thickness, to be enabled to surmount the new obstacle”.³⁷

This idea of obstruction to flow by the large vessels held sway for many years. Dwight³⁸ reported three cases of Pott’s disease with a sharp aortic inflection, and

²⁹ H. Bouvier, *Leçons cliniques sur les maladies chroniques de l'appareil locomoteur*, Paris, J.-B. Baillière, 1858, pp. 447–457.

³⁰ Bampfield, op. cit., note 24 above.

³¹ Morgagni, op. cit., note 16 above.

³² Jones, op. cit., note 23 above, p. 145.

³³ J. S. Donner, *Commentationis medico-chirurgicae de gibbositate*, pars 1, [M.D. Thesis], Göttingen, J. C. Dieterich, 1784, pp. 19–21.

³⁴ W. Vrolik, *Dissertatio anatomico-pathologia de mutato vasorum sanguiferorum decursu in scoliosi et cyphosi*, [M.D. Thesis], Amsterdam, C. A. Spin, 1823, pp. 12–13.

³⁵ J. R. Guérin, *Vues générales sur l'étude scientifique et pratique des difformités du système osseux, exposées à l'ouverture des Conférences Cliniques sur les difformités à l'Hôpital des Enfants de Paris, le 7 Aout 1839; suivies du résumé général de la première série des conférences cliniques*, Paris, Bureau de la Gazette Médicale, 1840, pp. 26–27.

³⁶ J. N. Corvisart, *Essai sur les maladies et les lésions organiques du coeur et des gros vaisseaux; extrait des leçons cliniques*, Paris, Migneret, 1806, pp. 114–117.

³⁷ E. Harrison, ‘Remarks upon the different appearances of the back, breast, and ribs in persons affected with spinal diseases: and on the effects of spinal distortion on the sanguineous circulation’, *Lond. med. phys. J.*, 1820, 44: 365–378.

³⁸ T. Dwight, ‘Distortion of the aorta in Pott’s disease’, *Am. J. med. Sci.*, 1900, 120: 429–435.

blamed frequent sudden death on “closure of the aorta” at this point. Klawansky³⁹ also reported a thirty-five-year-old kyphoscoliotic with acute angulation of his aorta, which he considered to be a mechanical obstruction to flow causing left ventricular hypertrophy, in an analogous way to aortic atresia. While very acute angulation of the aorta does occur in Pott’s disease of the spine, it probably does not cause significant obstruction to blood flow in other conditions. In the pulmonary circulation, too, Lewis, Davies, Samuels, and Hecht⁴⁰ could find no pressure change on passing a catheter up and down the main arteries.

In contrast to these views, Bampffield,⁴¹ although admitting that the circulation may be “deranged” and the subject liable to palpitations, stated that the heart was not subject to organic disease. Shaw, too, felt that the heart was much less commonly affected in spinal curvatures than was generally thought at the time.⁴² This was backed up by Hunter, who reported that the heart was completely normal in the autopsy of a woman of eighty with a severe kyphosis.⁴³

These views were not shared in France. Delpech,⁴⁴ from Montpellier, published his *Traité d’orthomorphie* in 1828, in which he emphasized that scoliosis, in contrast to kyphosis, could seriously affect the internal organs even though the deformity was slight. He mentioned an eighteen-year-old girl whose severe hypertrophy of her heart, due to the deformity, was the gravest aspect of her illness. However, he put forward no reasons for the hypertrophy, although he blamed the recurrent nosebleeds of another scoliotic on the displacement of the heart and great vessels causing blood to be diverted up to the head.

Forget, in a lucid but neglected paper published in 1836, described the terminal illness of a thirty-six-year-old gibbous patient.⁴⁵ He regarded the small lungs as the primary cause of the disordered function, and stated that it was the increased efforts of the right ventricle projecting the blood across them that led to its hypertrophy. Rokitansky⁴⁶ put forward the same idea in his famous *Handbuch der speciellen pathologischen Anatomie*. He attributed the enlargement of the right heart and “overfilling of the whole venous system” to the abnormal density of the lung tissue, which could become completely collapsed and airless in parts.

An intermediate position was taken by Latham, the famous cardiologist from St. Bartholomew’s Hospital, who wrote: “In almost all cases, where life continues with extreme deformity of the chest, the organic unsoundness of the parts within is complex. The lungs and the heart suffer equally; and, beside the common causes conducive to

³⁹ G. Klawansky, ‘Hochgradige mechanische Aortenstenose durch Kyphoskoliose’, *Klin. Wschr.*, 1925, 4: 831–832.

⁴⁰ C. S. Lewis, M. C. Davies, A. J. Samuels and H. H. Hecht, ‘Cor pulmonale (pulmono-cardiac syndrome). A case report’, *Dis. Chest*, 1952, 22: 261–268.

⁴¹ Bampffield, *op. cit.*, note 24 above.

⁴² J. Shaw, *On the nature and treatment of the distortions to which the spine, and the bones of the chest, are subject: with an enquiry into the merits of the several modes of practice which have hitherto been followed in the treatment of distortions*, London, Longman, Hurst, Rees, Orme, Brown & Green, 1823, p. 142.

⁴³ R. Hunter, ‘Notes of a dissection of a hunchback’, *Lond. med. Gaz.*, 1839, 24: 919–922.

⁴⁴ Delpech, *op. cit.*, note 26 above.

⁴⁵ Forget, *op. cit.*, note 27 above.

⁴⁶ C. Rokitansky, *Handbuch der Speciellen Pathologischen Anatomie*, Vienna, Braumüller & Seidel, 1844, vol. 2, p. 274.

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the unsoundness of both alike, each is continually helping in the unsoundness of the other.”⁴⁷

Later authors, such as Dusch,⁴⁸ and Traube,⁴⁹ emphasized the right ventricular hypertrophy. Paul, in a chapter entitled ‘Le coeur des bossus’ in his well-known textbook, recorded the dimensions of the heart in twenty cases of spinal deformity. He noted that the right ventricle could enlarge sufficiently to cause tricuspid incompetence and that the pulmonary second sound could be as loud as the aortic.⁵⁰

Soon afterwards, in 1899, Bachmann published an extensive pathological study.⁵¹ He reported a series of 197 kyphoscoliotic post mortems from Stuttgart and a further seventy-one from the literature. Bachmann detected hypertrophy of the right ventricle in 56.4 per cent, of the left in 17.5 per cent, and of both in 25.9 per cent. Associated with the left ventricular hypertrophy was the frequent finding of atheroma. This was also confirmed by Hirsch⁵² a year later. However, one of his seven autopsy subjects had a shrunken kidney which may have caused hypertension. Much later, in 1933, Dumas⁵³ reported two cases with hypertension, left ventricular hypertrophy and normal kidneys, and attributed the hypertension to an irritative effect of the deformity on the nerves to the heart.

Bachmann’s⁵⁴ finding that 82.3 per cent of his autopsies showed right ventricular hypertrophy confirmed in a large series what had previously been suggested from single cases or short series of cases. Within the next few years Romberg⁵⁵ and Fischer⁵⁶ published similar findings, and the former recollected that twenty-six out of thirty-eight high-grade scoliotics dying in Curschmann’s clinic at Leipzig died of “heart weakness”. The average age at death of fifteen men was forty years, and of eleven women was fifty-five years.

The remarkable absence of almost any significant contribution in the English language since Latham’s comments in 1846⁵⁷ was broken by the appearance in 1921 of an article by a Canadian, Finley.⁵⁸ He reported four cases of spinal deformity, two of whom died in right heart failure and were shown to have right ventricular hypertrophy. He attributed this, like Forget, to the increase in pulmonary vascular

⁴⁷ P. M. Latham, *Lectures on subjects connected with clinical medicine, comprising diseases of the heart*, 2nd ed., London, Longman, Brown, Green & Longmans, 1846, vol. 2, pp. 219–224.

⁴⁸ T. von Dusch, *Lehrbuch der Herzkrankheiten*, Leipzig, W. Engelmann, 1868, p. 113.

⁴⁹ L. Traube, ‘Ein Fall von Dilatation und Hypertrophie des rechten Ventrikels bei einem mit hochgradiger Scoliose und Verbildung des Brustkorbes behafteten Individuum’, *Ges. Beit. Path. Phys.*, 1878, 3: 354–358.

⁵⁰ C. Paul, *Diagnostic et traitement des maladies du coeur*, Paris, Asselin, 1883, pp. 519–524.

⁵¹ M. Bachmann, ‘Die Veränderungen der inneren Organe bei Hochgradigen Skoliosen und Kyphoskoliosen’, *Bibliotheca Medica*, 1899, Abt. D1, Heft 4.

⁵² C. Hirsch, ‘Ueber die Beziehungen zwischen dem Herzmuskel und der Körpermuskulatur und über sein Verhalten bei Herzhypertrophie’, *Dt. Arch. klin. Med.*, 1900, 68: 321–342.

⁵³ M. A. Dumas, ‘L’hypertension des bossus et des scoliotiques’, *Lyon méd.*, 1933, 151: 134–139.

⁵⁴ Bachmann, *op. cit.*, note 51 above.

⁵⁵ E. Romberg, *Lehrbuch der Krankheiten des Herzens und der Blutgefäße*, Stuttgart, F. Enke, 1906.

⁵⁶ W. Fischer, ‘Über die Sklerose der Lungenarterien und ihre Entstehung’, *Dt. Arch. klin. Med.*, 1909, 97: 230–251.

⁵⁷ Latham, *op. cit.*, note 47 above.

⁵⁸ F. G. Finley, ‘Spinal deformity as a cause of cardiac hypertrophy and dilatation’, *Can. med. Ass. J.*, 1921, 11: 719–723.

resistance which was due to the compression of the lungs. He felt that in the early stages of spinal deformities the dyspnoea was primarily due to the lungs, and in the later stages to the heart.

Finley's article was soon followed by three others describing right ventricular hypertrophy.^{59,60,61} Coombs⁶¹ felt that the small size of the lungs and the restricted diaphragmatic movement in kyphoscoliotics were indications for operating on the spine early in order to prevent the deformity.

The mainstream of thought at this time on the Continent followed that of the Frenchmen Barié⁶² and Huchard.⁶³ Both felt that the lungs were primarily at fault. Huchard argued for the term "poumon des bossus" rather than "coeur des bossus", which had been introduced by Paul in 1883.⁶⁴ Barié, however, was the first to state explicitly that it was the rise in pulmonary artery pressure that gave rise to the right ventricular hypertrophy. However, Dedic⁶⁵ postulated that the heart failure occurred because the respiratory efforts were insufficient to generate the negative pressure necessary to draw blood across the right heart. He felt this was especially so during exercise and was the cause of exertional dyspnoea. This theory has not been confirmed.

CAUSES OF DEATH

Although the cause of death was documented in isolated cases by Morgagni,⁶⁶ Bampfield,⁶⁷ and others, Stoll was the first to study this aspect in any detail. Describing his experience in Vienna in 1775, he wrote that "many of the hunchbacks perished of phthisis, peripneumonia, asthma, or hydrops of the chest".⁶⁸ Delpech⁶⁹ in France and Ward⁷⁰ in England also agreed that tuberculosis was a frequent finding in scoliotics, and Bachmann found evidence of it in 28.3 per cent of 197 autopsied kyphoscoliotics.⁷¹

In contrast Rokitansky commented that these subjects have an immunity to pulmonary tuberculosis.⁷² Bouvier⁷³ also denied any susceptibility to pulmonary tuberculosis although he conceded that scoliotics were particularly liable to con-

⁵⁹ E. P. Boas, 'The cardiovascular complications of kyphoscoliosis with report of a case of paroxysmal auricular fibrillation in a patient with a severe scoliosis', *Amer. J. med. Sci.*, 1923, 166: 89-95.

⁶⁰ W. D. Reid, 'Spinal deformity as a cause of cardiac hypertrophy', *J. Amer. med. Assoc.*, 1930, 94: 483.

⁶¹ C. F. Coombs, 'Fatal cardiac failure occurring in persons with angular deformity of the spine', *Br. J. Surg.*, 1930, 18: 326-328.

⁶² M. E. Barié, 'Le coeur dans les deviations du rachis et dans les déformations thoraciques', *Semaine méd.*, 1904, 24: 65-67.

⁶³ H. Huchard, *Maladies de l'appareil digestif et du l' appareil respiratoire*, Paris, J.-B. Baillière, 1911, pp. 107-114.

⁶⁴ Paul, op. cit., note 50 above.

⁶⁵ S. Dedic, 'La cardiopathie cyphotique', *Arch. mal. coeur vaiss. sang.*, 1930, 23: 33-36.

⁶⁶ Morgagni, op. cit., note 16 above.

⁶⁷ Bampfield, op. cit., note 24 above.

⁶⁸ M. Stoll, *Médecine pratique. Traduction nouvelle*, Paris, J.-A. Brosson, 1809, vol. 1, p. 191.

⁶⁹ Delpech, op. cit., note 26 above.

⁷⁰ W. T. Ward, *Practical observations on distortions of the spine, chest, and limbs: together with remarks on paralytic and other diseases connected with impaired or defective motion*, London, H. Renshaw, 1840.

⁷¹ Bachmann, op. cit., note 51 above.

⁷² Rokitansky, op. cit., note 46 above.

⁷³ Bouvier, op. cit., note 29 above.

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gestive and inflammatory illnesses, asthma, and haemoptysis. Much later, in 1932, Caussade and Tardieu⁷⁴ reported a severe scoliotic who recovered from extensive tuberculosis; and the view of Rokitansky⁷⁵ was also supported by Flagstad and Kollman.⁷⁶ They pointed out that the collapsed apices and immobile chest in scoliosis was the recommended treatment of tuberculosis and felt that there was no increased liability to developing a severe infection.

In his M.D. Thesis of 1865, Sottas⁷⁷ presented a series of cases of gradually increasing severity, in which the symptoms of dyspnoea and palpitations merged into those of heart failure and, ultimately, death. He described four autopsies showing right ventricular hypertrophy and hyperaemic lungs. His interpretation was that the blood was obstructed by the "narrowness" of the circulation in the small deformed lungs and that hypertrophy of the right heart is normally just sufficient to maintain health. However, exercise, lung or bronchial diseases, or heart conditions (such as arrhythmias) may disturb this fragile equilibrium. He recommended digitalis, diuretics, and venesection as possible treatments.

Sottas' views on bronchitis had been antedated by Forget,⁷⁸ who, in 1836, stated that even a trivial infection could hinder the circulation or respiration sufficiently to lead to asphyxia, cyanosis, and oedema. De Vesian⁷⁹ supported them further. He distinguished clearly between death due to heart failure ("asystole") and that secondary to bronchitis ("asphyxia") though admitting that it was often difficult to separate the two clinically. He recommended oxygen therapy when "arterialization" of the blood was insufficient during an episode of bronchitis. He also pointed out the exact correlation between the size of the thorax and the lungs, and that the smaller the lungs were, the larger was the heart.

Marfan⁸⁰ reported the case of a thirty-five-year-old man who exhibited right and left ventricular hypertrophy and tricuspid incompetence. The patient died during an attack of bronchitis. Marfan, like Sottas,⁸¹ interpreted the hypertrophy of the heart as a compensation for the small lungs. He argued that the infection had led to a respiratory death before the heart had had time to fail sufficiently to be fatal.

At about this time it became apparent that, quite apart from left ventricular failure or the effect of the lungs on the right ventricle, cardio-respiratory failure could occur in scoliotics as a result of congenital heart disease. Chantreau⁸² first published a case report of a florist with a severe spinal curvature who died in heart failure.

⁷⁴ G. Caussade and A. Tardieu, 'Sur les troubles cardiopulmonaires des gibbeux et leurs rapports avec l'étiologie tuberculeuse', *J. Méd. franç.*, 1932, 21: 195-198.

⁷⁵ Rokitansky, *op. cit.*, note 46 above.

⁷⁶ A. E. Flagstad and S. Kollman, 'Vital capacity and muscle study in one hundred cases of scoliosis', *J. Bone Jt Surg.*, 1928, 10: 724-734.

⁷⁷ E. Sottas, *De l'influence des déviations vertébrales sur les fonctions de la respiration et de la circulation*, [M.D. Thesis], Paris, A. Parent, 1865.

⁷⁸ Forget, *op. cit.*, note 27 above.

⁷⁹ F. de Vesian, *Étude sur la pathologie des poumons et du coeur chez les bossus*, [M.D. Thesis], Paris, A. Parent, 1884.

⁸⁰ A. Marfan, 'Observation pour servir à l'étude du pronostic de la bronchite chez les bossus', *Archs. gén. Méd.*, 1884, 14: 347-355.

⁸¹ Sottas, *op. cit.*, note 77 above.

⁸² Chantreau, 'Gibbosités rachitiques. Accès ce dyspnée et de cyanose. Congestions pulmonaires. Persistance du trou de Botal. Mort. Autopsie', *Bull. Soc. Anat. Paris*, 1867, 12: 525-526.

Autopsy examination showed an atrial septal defect large enough to admit a goose feather. Charrin⁸³ gave an account of a nineteen-year-old man under the title 'De la maladie bleue'. His main pulmonary artery was represented only by a fibrous cord and he also had a large ventricular septal defect. He had a very marked scoliosis and his heart occupied most of the thorax.

LABORATORY INVESTIGATIONS

It is remarkable how little had been added by about 1930 to the clinical and morbid anatomical findings that were established by 1850. Few new concepts had been introduced and the problems presented by the scoliotic patients were still seen as those which had faced the late eighteenth- and early nineteenth-century physicians. However, during the next few years the introduction of investigative techniques that could be applied to the live patient brought a more dynamic approach to the causes and mechanisms of the cardio-respiratory disability.

Brugsch⁸⁴ studied a few patients radiographically, and Eckhardt⁸⁵ used contrast media to illustrate the abnormal anatomy, but Rösler⁸⁶ was the first to use X-rays systematically. He found that the heart shape was often "mitralized" but argued that the abnormal anatomy of the chest may have accounted for this finding in some cases. Edeiken⁸⁷ found the same radiological abnormality and that the electrical axis in the electrocardiogram was as often deviated to the left as to the right. However, Adorno and White⁸⁸ in a larger survey of one hundred asymptomatic patients with chest deformities, found a normal axis in eighty-one and right axis deviation in seventeen of the remaining nineteen.

The long-assumed diminution in lung volumes was now confirmed by the application of spirometry. In fact, long beforehand, in 1844, John Hutchinson⁸⁹ had written "The [vital] capacity of those who suffer from curvature of the spine is most remarkably small. One person was so low as 27 cubic inches, being the utmost quantity he could throw out of his chest by one full expiration." Some years later, Hutchinson⁹⁰ noted that the mobility of the chest was decreased and that the vital capacity (VC) may fall as low as twenty cubic inches. His work was translated into German in 1849, and five years later Schneevogt⁹¹ published the findings of 127 spirometric

⁸³ Charrin, 'De la maladie bleue', *Semaine méd.*, 1890, 10: 413.

⁸⁴ T. Brugsch, 'Ueber das Verhalten des Herzens bei Skoliose', *Munch. med. Wschr.*, 1910, 57, ii: 1734.

⁸⁵ H. Eckhardt, 'Untersuchungen über die Lage von Brust-und Baueingewerden bei hochgradiger kyphoskoliose', *Zt. Orth. Chir.*, 1927, 48: 125-135.

⁸⁶ H. Rösler, 'Zur röntgenologischen Beurteilung des Herzgefäßbildes bei Thoraxdeformitäten; [(Kyphe)-Skoliose, reine Kyphe, Trichterbrust]. Nebst Bemerkungen über den Ösophagusverlauf', *Dt. Arch. klin. Med.*, 1929, 164: 365-377.

⁸⁷ J. Edeiken, 'The effect of spinal deformities on the heart', *Amer. J. med. Sci.*, 1933, 186: 99-110.

⁸⁸ A. R. Adorno and R. D. White, 'Electrocardiographic study of deformity of the chest', *Amer. Ht J.*, 1945, 29: 440-448.

⁸⁹ J. Hutchinson, 'Contributions to vital statistics obtained by means of a pneumatic apparatus for valuing the respiratory powers with relation to health', *J. Stat. Soc. Lond.*, 1844, 7: 193-212.

⁹⁰ J. Hutchinson, 'Thorax', in *Cyclopaedia of anatomy and physiology*, edited by R. B. Todd, London, Longman, Brown, Green, & Longmans, 1852, vol. 4, p. 1079.

⁹¹ G. E. V. Schneevogt, 'Ueber den Praktischen werth des Spirometer', *Zt. rat. Med.*, 1854, 5: 9-28.

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investigations, including those of three scoliotics. Two of these had vital capacities only slightly below the values predicted from their height by Hutchinson, but in one it was markedly decreased.

These early findings remained completely unsupported until the work of Flagstad and Kollman, published in 1928.⁹² They showed that the vital capacity was unaffected by low thoracic and lumbar curves. The reduction in VC was greatest in mid and high thoracic curves when these were severe and the respiratory muscles weak.

Two years later Anthony,⁹³ using a hydrogen mixing technique, measured the total lung capacity, residual volume and functional residual capacity as well as the VC in an asymptomatic kyphotic. He showed that the inspiratory capacity and expiratory reserve volume, and hence the VC, were severely reduced. The functional residual capacity and the total lung capacity were also diminished, but the residual volume was increased.

However the first thorough physiological assessment was carried out in 1939 in America by Chapman, Dill, and Graybiel.⁹⁴ They studied twelve patients, one of whom had been reported previously.⁹⁵ Three of their subjects complained of episodes of sudden loss of consciousness. This had been previously noted by Bouvier in 1858⁹⁶ and Sottas in 1865,⁹⁷ but only two scoliotics with cardiac arrhythmias (other than in a terminal illness) have been recorded.^{98,99} Another patient of Chapman *et al.*¹⁰⁰ was less dyspnoeic when lying face down. The researchers offered no explanation for this, though similar patients were later described by Kerwin,¹⁰¹ and Abrahamson and Abrahamson.¹⁰²

Chapman *et al.*¹⁰³ found that the VC of nine scoliotics was decreased and ranged from 0.56 to 2.52 L. They interpreted the high ratio of residual volume to total lung capacity as indicating emphysema. They also showed that the arterial oxygen saturation and pCO₂ were usually normal at rest, and that the cardiac output and circulation times were normal. They concluded that the chief effect of the deformity is on the lungs and that "pulmono-cardiac failure", which they never defined clearly, was "not analogous to the usual cor pulmonale or to Ayerza's disease". They emphasized that once "pulmono-cardiac failure" had appeared the prognosis was poor, and this

⁹² Flagstad and Kollman, *op. cit.*, note 76 above.

⁹³ A. J. Anthony, 'Untersuchungen über Lungenvolumina und Lungenventilation', *Dt. Arch. klin. Med.*, 1930, 167: 129-176.

⁹⁴ E. M. Chapman, D. B. Dill and A. Graybiel, 'The decrease in functional capacity of the lungs and heart resulting from deformities of the chest: pulmonocardiac failure', *Medicine*, 1939, 18: 167-202.

⁹⁵ R. C. Cabot, 'The interesting result of severe deformity of the thorax, Case Records of the Massachusetts General Hospital no. 19432', *New Eng. J. Med.*, 1933, 209: 854-855.

⁹⁶ Bouvier, *op. cit.*, note 29 above.

⁹⁷ Sottas, *op. cit.*, note 77 above.

⁹⁸ *Ibid.*

⁹⁹ Boas, *op. cit.*, note 59 above.

¹⁰⁰ Chapman *et al.*, *op. cit.*, note 94 above.

¹⁰¹ A. J. Kerwin, 'Pulmonocardiac failure as a result of spinal deformity. Report of five cases', *Archs. intern. Med.*, 69: 560-572.

¹⁰² L. Abrahamson and M. L. Abrahamson, 'Pulmonocardiac failure from chest deformity: with report of a case', *Ir. J. med. Sci.*, 1949, 281: 227-230.

¹⁰³ Chapman *et al.*, *op. cit.*, note 94 above.

was later supported by Hertzog and Manz.¹⁰⁴

The concept of “pulmono-cardiac failure” was reiterated by several authors (e.g. Lewis *et al.*;¹⁰⁵ Fischer and Dolehide;¹⁰⁶ Woods;¹⁰⁷ Abrahamson¹⁰⁸) over the two decades following the paper of Chapman *et al.*¹⁰⁹ However, “pulmono-cardiac failure” was never clearly defined, and seemed to include a rapid worsening of dyspnoea, a tendency to faint, and right heart failure. In a similar manner, Hanley, Platts, Clifton, and Morris¹¹⁰ emphasized the peculiarity of right ventricular failure in spinal deformities with their term “heart failure of the hunchback”, which was almost a literal translation of Paul’s (1883)¹¹¹ title. Tourniaire, Tartulier, Deyrieux, and Montouchet¹¹² also used the phrase “le coeur des cyphoscoliotiques” to describe the state of the heart in these subjects.

The application of cardiac catheterization gradually clarified the position. Lewis *et al.*,¹¹³ and Schaub, Bühlmann, Kälin, and Wegmann¹¹⁴ demonstrated that pulmonary hypertension did occur in kyphoscoliosis. Lewis *et al.*,¹¹⁵ and Bergofsky, Turino and Fishman¹¹⁶ showed that Harrison’s (1820)¹¹⁷ old concept of obstruction of the large pulmonary arteries by their distortion was untrue. They could find no pressure change on passing the catheter up and down the pulmonary arteries and at post mortem examination no obstruction was demonstrable.

Bergofsky *et al.*,¹¹⁸ in their very important paper from Columbia University, also showed that the wedge pressure and cardiac output were normal. The former was later confirmed by Shannon, Riseborough, Laercio, and Kazemi¹¹⁹ and Davies and Reid.¹²⁰ Thus the cause of the pulmonary hypertension was localized to the pulmonary capillary bed, and the right ventricular hypertrophy in scoliosis shown to be completely analogous to that of chronic lung diseases.

¹⁰⁴ A. J. Hertzog and W. R. Manz, ‘Right-sided heart failure (cor pulmonale) caused by chest deformity’, *Amer. Ht J.*, 1943, 25: 399–403.

¹⁰⁵ Lewis *et al.*, *op. cit.*, note 40 above.

¹⁰⁶ J. W. Fischer and R. A. Dolehide, ‘Fatal cardiac failure in persons with thoracic deformities’, *Arch. intern. Med.*, 1954, 93: 687–697.

¹⁰⁷ J. W. Woods, ‘Pulmonocardiac failure due to thoracic deformities’, *N. Carol. med. J.*, 1956, 17: 504–507.

¹⁰⁸ M. L. Abrahamson, ‘Pulmonocardiac failure associated with deformity of the chest’, *Lancet*, 1959, i: 449–450.

¹⁰⁹ Chapman *et al.*, *op. cit.*, note 94 above.

¹¹⁰ T. Hanley, M. M. Platts, M. Clifton and T. L. Morris, ‘Heart failure of the hunchback’, *Quart. J. Med.*, 1958, 27: 155–171.

¹¹¹ Paul, *op. cit.*, note 50 above.

¹¹² A. Tourniaire, M. Tartulier, F. Deyrieux and M. Montouchet, ‘Le coeur pulmonaire chronique des cyphoscolioses’, *J. Méd. Lyon.*, 1962, 43: 1581–1607.

¹¹³ Lewis *et al.*, *op. cit.*, note 40 above.

¹¹⁴ F. Schaub, A. Bühlmann, R. Kälin and T. Wegmann, ‘Zur Klinik und Pathogenese des sogenannten Kyphoskolioseherzens’, *Schweiz. Med. Wschr.*, 1954, 84: 1147–1150.

¹¹⁵ Lewis *et al.*, *op. cit.*, note 40 above.

¹¹⁶ E. H. Bergofsky, G. M. Turino and A. P. Fishman, ‘Cardiorespiratory failure in kyphoscoliosis’, *Medicine*, 1959, 38: 263–317.

¹¹⁷ Harrison, *op. cit.*, note 37 above.

¹¹⁸ Bergofsky *et al.*, *op. cit.*, note 116 above.

¹¹⁹ D. C. Shannon, E. J. Riseborough, M. V. Laercio and H. Kazemi, ‘The distribution of abnormal lung function in kyphoscoliosis’, *J. Bone Jt Surg.*, 1970, 52A: 131–144.

¹²⁰ G. Davies and L. Reid, ‘Effect of scoliosis on growth of alveoli and pulmonary arteries and on right ventricle’, *Arch. Dis. Childh.*, 1971, 46: 623–632.

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During the 1950s the lung volumes of scoliotics were studied extensively. Most authors agreed that the vital capacity, residual volume, functional residual capacity, and total lung capacity were all reduced in adults,^{121,122} and even in adolescents.¹²³ Caro and Gucker were able to relate the small size of the lungs to the decreased compliance of the lungs and chest wall,¹²⁴ but the effect of spinal fusion on lung volumes is still not settled.¹²⁵

The idea that emphysema was common in scoliotics, originally expressed by Forget in 1836,¹²⁶ was propagated as late as 1955 when Larmi *et al.*¹²⁷ argued that the increased ratio of residual volume to total lung capacity was due to emphysema. A year later however, Bedell, Marshall, Du Bois, and Comroe¹²⁸ found similar lung volumes by plethysmographic and dilution techniques, thus excluding the possibility of gas being "trapped" in the lungs. Bergofsky *et al.*¹²⁹ then obtained normal results with single and multibreath nitrogen washout techniques, and Sadoul and Cherrier,¹³⁰ and Ting and Lyons¹³¹ found that the helium equilibration time in a closed circuit was also normal. These physiological studies were later backed up by histological evidence provided by Dunnill¹³² and Reid,¹³³ which also showed no evidence of emphysema.

Analysis of arterial blood samples was first systematically carried out by Schaub *et al.*¹³⁴ They detected hypoxaemia in the absence of a raised pCO₂ in some patients. Since then, the use of radioactive xenon isotopes has confirmed that the distribution of blood flow to the lungs is abnormal in scoliotics.¹³⁵ Hypoxaemia is therefore more often due to suboptimal matching of ventilation and perfusion than to hypoventilation.

This mismatching of ventilation and perfusion persists during exercise¹³⁶ and results

¹²¹ F. Schaub, A. Bühlmann and R. Kälin, 'Das "Kyphoskolioseherz" und seine Pathogenese', *Cardiologia*, 1954, 25: 147-152.

¹²² T. K. I. Larmi, J. Päätilä and M. J. Karvonen, 'Studies of pulmonary function in kyphoscoliosis after tuberculous spondylitis', *Ann. Med. Int. Fenn.*, 1955, 44: 57-69.

¹²³ A. Bühlmann and W. Gierhake, 'Die Lungenfunktion bei der jugendlichen Kyphoskoliose', *Schweiz. med. Wschr.*, 1960, 90: 1153-1155.

¹²⁴ C. G. Caro and T. Gucker, III, 'Effects of kyphoscoliosis upon mechanics of breathing in children', *Amer. Rev. Tuberc.* 1958, 78: 326.

¹²⁵ P. A. Zorab, 'The medical aspects of scoliosis', in *Scoliosis*, 2nd ed., edited by J. I. P. James, Edinburgh, Churchill Livingstone, 1976, pp. 334-343.

¹²⁶ Forget, *op. cit.*, note 27 above.

¹²⁷ Larmi *et al.*, *op. cit.*, note 122 above.

¹²⁸ G. N. Bedell, R. Marshall, A. B. DuBois and J. H. Comroe jr., 'Plethysmographic determination of the volume of gas trapped in the lungs', *J. clin. Invest.*, 1956, 35: 664-670.

¹²⁹ Bergofsky *et al.*, *op. cit.*, note 116 above.

¹³⁰ P. Sadoul and F. Cherrier, 'L'insuffisance respiratoire des gibbeux', *J. franç. Méd. Chir. Thorac.*, 1963, 17: 167-180.

¹³¹ F. Y. Ting and H. A. Lyons, 'The relation of pressure and volume of the total respiratory system and its components in kyphoscoliosis', *Amer. Rev. resp. Dis.*, 1964, 89: 379-386.

¹³² M. S. Dunnill, 'Quantitative observations on the anatomy of chronic non-specific lung disease', *Med. Thoracal.*, 1965, 22: 261-274.

¹³³ L. Reid, 'Autopsy studies of the lungs in kyphoscoliosis', in *Proceedings of a symposium on scoliosis*, edited by P. A. Zorab, London, National Fund for Research into Poliomyelitis and other Crippling Diseases, 1965, pp. 71-78.

¹³⁴ Schaub *et al.*, *op. cit.*, note 121 above.

¹³⁵ C. T. Dollery, P. M. S. Gillam, P. Hugh-Jones and P. A. Zorab, 'Regional lung function in kyphoscoliosis', *Thorax*, 1965, 20: 175-181.

¹³⁶ J. M. Shneerson, 'Cardio-respiratory function in thoracic scoliosis', D.M. Thesis, University of Oxford, 1976.

in an abnormally low PaO₂. During exercise the pulmonary artery pressure and respiratory rate rise quickly and the maximum oxygen uptake is diminished. These abnormalities are proportional to the vital capacity.¹⁸⁷ This therefore plays a central role in determining the symptoms of scoliotics and probably their likelihood of developing right ventricular hypertrophy.

SUMMARY

Hunchbacks have always been physically conspicuous and it is therefore not surprising that their cardio-respiratory problems attracted attention from the earliest times. Hippocrates discussed their shortness of breath in some detail, but little further advance was made until the eighteenth century. Morbid anatomical studies by Morgagni (1769) and others demonstrated the distortions of the intrathoracic structures, but various views on the relative importance of the pulmonary, cardiac, and arterial derangements were held. During the nineteenth century the natural history of a transition from breathlessness through a phase of cardiac failure to a premature death was clarified. Acute respiratory infections and congenital heart defects were also recognized as common causes of death.

Remarkably little conceptual progress was made between 1850 and 1950. Apart from the pioneer spirometric measurements of Hutchinson in 1844, the application of physiological investigations was also curiously delayed. However, since 1950 the small size of the lungs, the absence of emphysema, and poor matching of the distribution of ventilation and perfusion have been demonstrated. Cardiac catheterization has shown that pulmonary hypertension is common, especially on exercise, and that there is a close interrelationship between the cardiac and respiratory abnormalities in these subjects.

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¹⁸⁷ *Ibid.*