

# Prevalence of Scoliosis in Women with Visual Deficiency

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**Abstract.** Whether visual impairment influences the prevalence of scoliosis in humans or not remains controversial. The purpose of this study was to assess the prevalence of scoliosis in blind women in a Mediterranean region.

**Material and Method:** A total of 26 blind women aged 40 years (median, range 20 - 67) were screened for scoliosis. The existence of a possible trunk hump was measured by the forward bending test using the Pruijjs Scoliometer. Reading of an Angle of Trunk Inclination (ATI) greater or equal to 7 degrees was used as a cut-off criterion for radiological examination. Standing postero-anterior and lateral spinal radiographs were obtained. Menarche and circadian rhythm was recorded.

**Results :** 11 out of 26 women had a scoliosis with an average Cobb angle of 19 degrees (range 12 - 28). The average ATI was 8 degrees. Thoracolumbar was the most common type of curve identified (9 out of 11, 6 were to the right and 3 to the left). The median age of menarche was 13 years (range, 11 - 15). None of the blind women reported any difficulty sleeping and had a circadian rhythm related to a 24-hour day.

**Discussion:** The prevalence of scoliosis was 42.3%, while the prevalence in the general population in the same regime is 2.9%. Blind women had a later age of menarche (13 versus 12.58 yrs) compared with normal girls. The postural etiology of scoliosis in blind people has been reported. The possible role of light in association to melatonin production, age at menarche and high prevalence of scoliosis in blind women is presented and discussed.

**Keywords:** idiopathic scoliosis, blindness, prevalence of IS, menarche, melatonin

## 1. Introduction

The cause of adolescent idiopathic scoliosis (AIS) in humans remains obscure. A wide range of aetiological causes have been proposed including neuromuscular abnormalities, connective tissue anomalies, vestibular dysfunction, melatonin secretion disturbance, platelet microstructure anomalies, mechanical factors, growth related and developmental problems, asymmetry of the brainstem, genetic factors, equilibrium dysfunctions and impairment of proprioception. These concepts have led to the idea that a disturbance of postural control is involved but no single factor has been identified so far. Many authors consider that a relationship exists between the origin of scoliosis and problems of balance in the patients. Visual impairment has been shown to increase the prevalence of idiopathic scoliosis in human subjects when compared to the prevalence of the general population [1-6].

The objective of this study was to select data from scoliosis screening in an Institute of blind women in a Mediterranean region and study the prevalence of scoliosis in relation to the age at menarche, the lack of light stimulus and the possible role of melatonin.

## **2. Material and Method**

### *2.1. The Study Population*

A total of 26 blind women in an Institute for blind people with median aged 40 years (range 20 – 67 years) were screened for scoliosis. The causal pathologies for the blindness were various: retina or refraction problems, crystalline lens abnormalities, or oculomotor pathology. The patients with general neurological pathology were excluded.

### *2.2. The Measurements*

The presence of a trunk hump was elicited by the forward bending test applying the Pruijjs Scoliometer. An Angle of Trunk Inclination (ATI) greater or equal to 7 degrees was used as a cut-off criterion for radiological examination. For positive cases, a standing posteroanterior and lateral spinal radiograph was obtained. The Cobb angle was measured and a curve of 10 degrees or more was used to define a scoliosis following SRS guidelines.

### *2.3. The Age at Menarche and Circadian Rhythm*

The blind women were questioned about their menarche and their circadian rhythm was also recorded.

## **3. Results**

### *3.1. The Measurements*

11 out of 26 women recorded a scoliosis, with an average Cobb angle of 19 degrees (range 12 - 28). The average ATI was 8 degrees (range 7 to 10 degrees). Thoracolumbar curves were the most common type identified ( 9 out of 11). 6 thoracolumbar curves were to the right and 3 were to the left. Of the remaining two cases, one had a lumbar curve to the right and the other a thoracic curve to the right.

### *3.2. The Age at Menarche and Circadian Rhythm*

The median age of menarche of the 26 blind women was 13 years (range, 11 - 15). All blind women reported no difficulty in sleeping and had a normal 24-hour day circadian rhythm.

#### 4. Discussion

The present study has demonstrated a prevalence of scoliosis in women with visual impairment as 42.3% against that of the general population in Greece of 2.9% [7].

##### *4.1. The Postural Aetiology of Scoliosis in People with Visual Impairment*

Vision by its function of environment exploration contributes to the postural control of the trunk. There is an association between idiopathic scoliosis and postural control in which vision is involved. Ponseti et al [8] found ocular problems in 32 patients with idiopathic scoliosis. In their study, the severity of the ocular lesions was related to the severity of the idiopathic scoliosis. Oculomotor modifications have also been found in association with idiopathic scoliosis. Close connections between the anomalies of the propio-oculo-labyrinthin and the scoliosis have been reported [9]. A genetic cause for posterior basicanium (PB) asymmetry and significant semi-circular canal malformation has also been reported in people with AIS [10]. Several studies report a family association of complete horizontal gaze palsy and severe progressive scoliosis [11]. In the series of Catanzariti et al [6] 18 out of 26 children who were visually impaired had a non-evolving three-dimensional scoliosis.

##### *4.2. The Role of Melatonin in the Aetiology of Scoliosis in People with Visual Impairment*

Data relating to scoliosis and melatonin in humans is relatively sparse and that already published does not clearly support a role for melatonin producing scoliosis in humans. The observation that experimental pinealectomy and a deficiency of melatonin (the principal product of the pineal gland in newborn chickens) leads to a spinal deformity similar to idiopathic scoliosis in man [12-14] lead to the development of a new neuroendocrine hypothesis for the cause of idiopathic scoliosis. However, no alteration in serum or urinary melatonin in patients with AIS has been recorded [15-18]. Moreover, an increased incidence of scoliosis has not been observed in children after pinealectomy or pineal irradiation because of pineal neoplasias, although they have may lack serum melatonin [19,20]. Thus the data regarding human melatonin levels is mixed at best and the melatonin deficiency as a causative factor in the aetiology of AIS cannot be supported.

##### *4.3. Hypothesis*

Melatonin is the main mediator that transfers changes in environmental light to human cells. Thus the diurnal variation of melatonin is due to environmental light conditions. Melatonin production is stimulated by darkness. Under conditions of darkness melatonin production increases and light reduces the melatonin production [21,22]. Bellastella et al [23] studied a group of 11 totally and 16 partially blind adult patients and found elevated melatonin levels both in patients with total blindness and in those with light perception compared to controls. Erren in 2002 [24] suggested that a lower incidence of cancer in blind people may be due to greater melatonin production.

The results of the current study suggest that melatonin might be involved in the pathogenesis of human idiopathic scoliosis in a way that could explain the higher prevalence of scoliosis in people with visual impairment. Melatonin overproduction due to lack of light stimulus in these people leads to late menarche and longer exposure to possible coexisting detrimental factors in the pathogenesis of AIS.

#### 4.4. Melatonin and Age at Menarche

A controversy exists whether blind girls with no light stimuli experience delayed puberty. Zacharias in 1964 [25] reported an earlier age at menarche while Thomas in 1967 [26] reported no difference and Segos in 1995 [27] noted blind girls have a later age of menarche. Jafarey et al in 1971 [28] reported that artificial lighting results in a decrease in the age of menarche. Retinal responses to environmental lighting mediate an expanding list of neuroendocrine effects, including control of pubescence, ovulation, and a large number of daily rhythms [29]. Sexual maturation can be delayed in experimental animals by exogenous melatonin administration or by short day exposure [30,31]. Animals kept in constant darkness show a delay in sexual maturation. However, the neuroendocrine pathways, which mediate the effects of blindness upon human sexual development, are not completely understood. Melatonin acts on the gonads indirectly, reducing the secretion of gonatotropines and mainly that of LH. Blind women presented late age at menarche (median age 13 yrs) in this study, as opposed to non-blind individuals (mean age 12,58 years in Greece). In view of these results, it is hypothesized that blindness and the delay sexual maturation might render blind women to longer exposure to the detrimental causative factors of AIS, leading to the increased reported prevalence of the condition.

## References

- [1] A.E. Geissele, M.J. Kransdorf, C.A. Geyer, J.S. Jelinek, B.E. Van Dam. Magnetic resonance imaging of the brain stem in adolescent idiopathic scoliosis. *Spine* **16** (1991), 761-3.
- [2] K.L.Kesling ,K.A. Reinker . Scoliosis in twins: a meta-analysis of the literature and report of six cases. *Spine* **22** (1997), 2009-14.
- [3] K.Yamada , H.Yamamoto ,Y. Nakagawa , et al. Etiology of idiopathic scoliosis. *Clin Orthop* **184** (1984), 50-7.
- [4] R.L. Barrack , T.S. Whitecloud ,S.W. Burke ,S.D. Cook ,A.F. Harding . Proprioception in idiopathic scoliosis. *Spine* **9** (1984), 681-5.
- [5] R.G.Burwell . The proprioceptive spinocerebellar loop concept for the aetiology of adolescent idiopathic scoliosis. *J Bone Joint Surg [Br]* **76 (Suppl1)** (1994), 19
- [6] J. Catanzariti , E. Salomez, J.M. Bruandet, A. Thevenon. Visual Deficiency and Scoliosis *Spine* **26 (1)** (2001), 48–52
- [7] T.B. Grivas , K. Koukos , U.I.Koukou , C. Maziotou , B.D.Polyzois .The incidence of idiopathic scoliosis in Greece--analysis of domestic school screening programs. *Stud Health Technol Inform.* **91** (2002),71-5.
- [8]. V. Ponseti , G.K.Von Noorden , H.M.Burian . Anterior chamber angle of the eye in patients with idiopathic scoliosis. *J Bone Joint Surg [Am]* **42** (1960),1448.
- [9] J.P. Deroubaix, D. Rousie. Accurate vestibular assessment : interest in the Adolescent Idiopathic Scoliosis. 11th Zorab Symposium, 2006
- [10] D. Rousie, O. Joly, P. Salvetti, J.P. Deroubaix, A. Berthoz. Idiopathic Scoliosis and the Basicranium Assymetry. 11th Zorab Symposium, 2006

- [11] E.K. Dretakis , N. Panos , M.D. Kondiyannis . Congenital scoliosis associated with encephalopathy in five children of two families. *J Bone Joint Surg [Am]* **56** (1974),1747–50.
- [12] M. Machida , J. Dubousset , Y. Imamira , T. Iwaya , T. Yamada , J. Kimura . An experimental study in chickens for the pathogenesis of idiopathic scoliosis. *Spine* **18** (1993), 1609-13.
- [13] X. Wang , H. Jiang , J. Raso , et al. Characterisation of the scoliosis that develops after pinealectomy in the chicken and comparison with adolescent idiopathic scoliosis in humans. *Spine* **22** (1997), 2626-35.
- [14] M. Machida , J. Dubousset , Y. Imamura , et al. Melatonin: a possible role in pathogenesis of adolescent idiopathic scoliosis. *Spine* **21** (1996), 1147-52.
- [15] A.B. Fagan , D.J. Kennaway , A.D. Sutherland . Total 24-hour melatonin secretion in adolescent idiopathic scoliosis: a case-control study. *Spine* **23** (1998),41-6.
- [16] A.S.Hilibrand , L.C. Blakemore , R.T. Loder, M.L. Greenfield, F.A. Farley, R.N. Hensinger, M. Hariharan. The Role of Melatonin in the Pathogenesis of Adolescent Idiopathic Scoliosis. *Spine* **21(10)** (1996), 1140-1146.
- [17] K.M.Bagnall ,V.J. Raso , D.L.Hill , et al. Melatonin levels in idiopathic scoliosis: diurnal and nocturnal serum melatonin levels in girls with adolescent idiopathic scoliosis. *Spine* **21** (1996), 1974-8.
- [18] W. Brodner , P. Krepler, M. Nicolakis, M. Langer, A. Kaider, W. Lack, F. Waldhauser. Melatonin and adolescent idiopathic scoliosis. *J Bone Joint Surg [Br]* **82** (2000), 399-403.
- [19] A. Etzioni , R. Luboshitzky , D. Tiosano , et al. Melatonin replacement corrects sleep disturbances in a child with pineal tumor. *Neurology* **46** (1996), 261-3.
- [20] J. Murata , Y. Sawamura , J. Ikeda , S. Hashimoto ,K. Honma . Twenty four hour rhythm of melatonin in patients with a history of pineal and/ or hypothalamo-neurohypophyseal germinoma. *J Pineal Res* **25** (1998),159-66.
- [21] M.L. Batrinos . *Endocrinology*. Athens, Greece: P.Paschalidis Medical Buplications, 1995.
- [22] D. Aravantinos . *Physiology of the woman*. Athens, Greece: Parizianos Publications, 1985.
- [23] A. Bellastella , A.A. Sinisi ,T. Criscuolo , A. De Bellis , C. Carella , S. Iorio, A.M. Sinisi , F. Parlato , T. Venditto , G. Pisano . Melatonin and the pituitary-thyroid axis status in blind adults: a possible resetting after puberty. *Clin Endocrinol (Oxf)* **43(6)** (1995), 707-11.
- [24] T.C. Erren , R.G. Stevens . Light , melatonin and internal cancers-recent facts and research perspectives. *Gesundheitswesen* **64** (2002), 278-283.
- [25] L. Zacharias , R.J. Wurtman . Blindness: its relation to age at menarche. *Science* **144** (1964), 1154.
- [26] J.B. Thomas , D.J. Pizzarello . Blindness, biologic rhythms, and menarche. *Obstet Gynecol* **30(4)** (1967), 507-9.
- [27] C. Segos . Puberty. In: Michalakis SP, Goumalatsios NG editors. *Honorary volume for Professor DJ Aravantinos*, Athens, Greece: Parisianos Publications, 1999;629–633.
- [28] N.A. Jafarey , M.Y. Khan , S.N. Jafarey . Effect of artificial lighting on the age of menarche. *Lancet* **3;1(7701)** (1971), 707.
- [29] R.J.Wurtman ,R.N. Neer . Good light and bad. *New England J Medicine* **282(7)** (1970), 394-395.
- [30] J. Arendt . Melatonin. *Clinical Endocrinology* **29** (1988),205-29.
- [31] R.D. Utiger . Melatonin: The hormone of darkness. *New Eng J Med* **327** (1992), 1377-9.

# Patterns of Extra-Spinal Left-Right Skeletal Asymmetries in Adolescent Girls with Lower Spine Scoliosis: Relative Lengthening of the Ilium on the Curve Concavity & of Right Lower Limb Segments

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**Abstract.** Extra-spinal skeletal length asymmetry have been reported for the upper limbs and periapical ribs of patients with thoracic adolescent idiopathic scoliosis. This paper reports (1) a third pattern with relative lengthening of the ilium on the concavity of *lower spine scolioses*, and (2) a fourth pattern of relative lengthening of the right total leg and right tibia unrelated statistically to the severity or side of *lower spinal scolioses*. The findings pose the question: are these anomalous extra-spinal left-right skeletal length asymmetries unconnected with the pathogenesis of AIS. Or, are they indicative of what may also be happening to some vertebral physes as an initiating pathogenic mechanism for the scoliosis?

**Key words:** Scoliosis, etiology, bilateral symmetry, pelvis, legs

## 1. General introduction

An anomalous feature detected in girls with AIS is extra-spinal left-right skeletal length asymmetries (Figure 1). Upper limb length asymmetry with relative lengthening on the convexity of right thoracic and thoracolumbar AIS curves was reported in clinic patients [1-3], in preoperative patients [4], and very recently in school screening referrals [5]. A second site reported was periapical rib lengthening on the concavity of right thoracic AIS in girls (RT AIS-F)[6]. Together with other clinical as much experimental evidence the periapical rib finding was used to assemble a thoracospinal concept of etiopathogenesis for the initiation of RT AIS-F [7]. Subsequent research using radiological and mathematical techniques did not confirm the rib findings [8,9].