

MORQUIO SYNDROME – A CASE STUDY IN AYURVEDIC PERSPECTIVE

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ABSTRACT

Kubjata is one of the *Vataja Nanatmaja vikara*; this comes under the umbrella of *Janmabala pravrutta vyadhi*. Here is a case of *Kubjata* compared to Morquio Syndrome. Morquio syndrome is a rare inherited autosomal recessive disorder characterized by the accumulation of mucopolysaccharides (Glycosaminoglycans) in various body tissues. It is rare cause of dwarfism. In this case the *Shaman* line of management proves best to conquer the symptoms, improve quality of life and to delay prognosis of the disease.

Keywords: Morquio syndrome, *Kubjata*, *Janmabala pravrutta vyadhi*, *Laksha*, *Mahisha dravaka*, *Bhargavaprokta rasayana*.

INTRODUCTION

Morquio syndrome or MPS IVA is a rare type of autosomal recessive mucopolysaccharide storage disease which is characterized by the inability to metabolize keratin sulphate. Mucopolysaccharide which is also known as Glycosaminoglycans (GAGs), are long-chained carbohydrates vital for the formation of bone, cartilage, tendons, corneas, skin, and connective tissue¹. The deficiency of lysosomal enzymes required for the metabolism of the various mucopolysaccharides results in the accumulation of the respective substrate in the cells, leading to progressive cellular damage and clinic- pathological changes.

It was first described in 1929 by Luis Morquio in Montevideo, Uruguay². The symptoms associated with Morquio's syndrome are usually noticed between one to three years of age and can include abnormal heart development, abnormal skeletal development, hyper-mobile joints, large fingers, knock-knees, widely spaced teeth, a bell-shaped chest, compression of the spinal cord, an enlarged heart and dwarfism. The syndrome is estimated to occur in one in every 200,000 births, with a family history of the syndrome raising one's risk of developing the condition³.

Diseases are classified basically as *Adibala pravrutta*, *Janma bala pravrutta*, *Doshabala pravrutta* and *Swabhava bala pravrutta* based on onset of the diseases. When the cause is before the birth, either it could be *Adi bala pravrutta* or *Janma bala pravrutta vyadhi*⁴. Clinical presentation at birth and later varies among different disorders that the person develops. Less height and deformities is characteristic feature of *Kubjata*. This case is about such a child with delayed milestone and skeletal deformity which is treated with the diagnosis as *Kubjata*.

CASE REPORT

An eight years old female, with normal intelligence who was born out of a non consanguineous marriage, had come to Shri Dharmastala Manjunatheshwara college of Ayurveda and Hospital, Udupi, India with a complaint of delayed linear growth, imbalance, difficulty in climbing the stairs & delayed milestones. Family history is free from any skeletal disorder. The physical appearance was characterized by short stature, short neck, protuberant chest, scoliosis, and waddling gait.

EXAMINATIONS

Her facial features presented frontal bossing, coarse facies with 1/3 of face prominent, low nasal bridge, and low set ears, thin lips with wide mouth and long philtrum. On oral examination, teeth with sharp cusps and rough enamel texture were found. Patient also presented bell

shaped chest with ribs flared out at the bottom, mild Clinodactyly of left and right little finger along with Brachydactyly, short neck (length 6 inches, Fraction- 7.8), Hallux varum, and was measuring 93cm, weighing 12kg. Medical examination of all major systems revealed no dysfunctional changes. Power of all the four limbs was 5/5. She can walk only for 20 meters as she develops pain in joints and exhaustion. Her intellectual functioning is normal.

INVESTIGATIONS:

The orthopedic and radiographic evaluation indicated gross skeletal deformity i.e, MRI suggestive of Hypo plastic L1 vertebral body causing Kyphotic deformity, Anterior vertebral body inter beaking T12 to L1, Mild acute Scoliosis at T12 – L1. The hand-wrist radiograph showed proximally narrow metacarpals, and an assay for Leukocytes galactose 6 sulfatase enzyme – 0.0nmol 17/hr/mg ; Urine MPS Electrophoresis – Band at the region of keratase sulfate; Leukocytes β - galactosides enzyme (MPS 10B) - 115.4 nmol/hr/mg.

A diagnosis of mucopolysaccharidosis type IVA was reached based on the clinical, dental, radiographic findings of the patient.

TREATMENT

As such there is no specific treatment mentioned separately in classics, the treatment was planned in the view to delay the prognosis of disease and to improve the quality of a life of a patient. Treatment given on admission is

Table 1: List of medications given on admission

Cap. <i>Laksha</i> _(SDM)	250mg tid
<i>Bargavaprokta Rasayana</i>	10gm tid
<i>Mahisha dravaka</i>	10ml tid
<i>Vestana</i> to both the lower and upper limbs with <i>maha masha tailam</i> .	

Table 2: List of medications on discharge

Cap. <i>Laksha</i> _(SDM)	250mg tid
<i>Bargavaprokta Rasayana</i>	10gm tid
<i>Mahisha dravaka</i>	10ml tid

IMPROVEMENT

After 2 months of treatment now patient is able to jump by herself without any imbalance. She is able to walk about 100meters without exertion. Stability of patient in terms of stamina to do activities improved.

FOLLOW UP TREATMENT

Same medication was continued with expectation of improvement in bone density, to maintain the body weight and to improve the quality of life of the patient.

DISCUSSION

Mucopolysaccharidosis (MPS) are a rare group of metabolic disorders due to deficiency of enzymes responsible for degradation of Glycosaminoglycans (dermatan sulfate, heparin sulfate, keratin sulfate). Based on clinical and radiological studies, these are grouped into six types. Mucopolysaccharidosis 4 is subdivided in two types. MPS 4A and MPS 4B, both are difficult to differentiate clinically⁵. They are spondyloepiphysio-metaphyseal dysplasia generally diagnosed in the second year of life when the child starts walking.

Samvahana of the body depends on *dhatu*. Irregularity in the body shape mainly because of malformation of *Asthi dhatu* and lean body stature indicates malformation of *Mamsa dhatu*. These two *dhatu* are essential for physical activities like walking, jumping, running etc. Since there is no evidence of appropriate etiology, a suspicion of *Janma bala pravrutta vyadhi* is made. *Kubjata* is one amongst *janmabala pravrutta vyadhi* and *vataja nanatmaja vikara*⁶. According to *acharya Madhavakara*, *Kubjata* refers to elevated chest or back⁷. *Acharya Vagbhata* explained it as *Avanama* ie bowing of the body⁸. Considering the subtypes of this condition is diagnosed as *Bahirayama Kubjata*⁹.

Since the child was able to walk at the beginning and presence of deformity rules out the possibility of *Phakka roga*. With due consideration of

Mamsa and *Asthi dhatu* involvement in this disease the treatment is planned. *Laksha*¹⁰ is helpful to correct the *vikruta Asthi dhatu*. *Mahisha dravaka* is a formulation adopted from *Arka prakasha*, which helps in correcting the *Mamsa dhatu*. *Bhargava prokta rasayana*¹¹ improves metabolism and proper development of the body.

CONCLUSION

There is no specific treatment for Morquio syndrome, hence it demands for an effective approach. The malformation of *Asthi* and *Mamsa dhatu* can be treated by *Laksha*, *Mahisha dravaka* and *Bhargava prokta rasayana* so as to improve quality of life, as evidenced in this case.

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