

## Neurocognitive Profile of Turner Syndrome

Turner syndrome (TS) is associated with a characteristic neurocognitive profile which includes average to low full scale intelligence quotient (FSIQ) with a significant discrepancy between verbal IQ (VIQ) and performance IQ (PIQ) [1]. They have average verbal skills but weaker non verbal skills like visuospatial, perceptual, processing speed and executive functioning leading to problems in academics, especially mathematics [2]. Mental retardation associated with a ring chromosome karyotype is rare (2-4%).

We assessed an adolescent girl with Turner syndrome in our learning disability clinic, referred for difficulties in comprehension, written expression and mathematics. Her scores on Wechsler Intelligence Scale for Children were VIQ-103, PIQ-99 with a Full Scale IQ-101 *i.e.* normal intelligence capacity (90-109). The verbal subtests implicated she had average abilities in verbal reasoning, a good working memory but possible deficits in abstract thinking. She had a good number sense but struggled with numerical sequencing and reasoning. Her performance subtests implied she was poor in areas of spatial visualization, visual perception and visuomotor coordination. In tests of Visuo Motor Integration, she was below average for speed and dexterity and tests of Visual Perception confirmed her poor visual working memory.

On the Woodcock Johnson–Revised Psychoeducational Battery, her scaled scores of broad reading, broad mathematics and broad written language were below average causing a significant potential - achievement discrepancy, the essential criteria for diagnosis of specific learning disabilities in areas of reading, writing and mathematics. In mathematics, there was difficulty with reasoning and applied problems skills. There was a slowed response on all timed tests *i.e.* reading, writing and mathematics fluency suggesting processing speed deficits. In lieu of above findings a diagnosis of generalized learning disability (LD) in areas of reading, writing and mathematics with Turner syndrome was made.

The etiology of impaired cognition in TS is multifactorial. Imaging studies have shown that the parieto-occipital cortex involved in visual-spatial processing is smaller in Turner syndrome. Estrogen deficiency influences memory, processing speed and motor skills which improves on supplementation but deficits in visual perceptual and spatial skills persist lifelong. Genetic studies have mapped the visuospatial deficits to the PAR 1 of Xp22.3 [3].

Research on neurocognitive profile has led to specific instructional recommendations for school aged girls to improve visuospatial, organizational skills to compensate for non verbal learning disabilities (NVLD). A full psychoeducational evaluation along with occupational therapy and physiotherapy is recommended. It is imperative to screen for dyslexia, present in our case; as it may be overlooked since TS association with NVLD is more known [2,5]. On detection of LD, remediation remains the cornerstone [5]. Academic provisions give the child an opportunity to continue in mainstream and be able to perform and compete with peers in schools.

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