TPM Vol. 32, No. S1, 2025
ISSN: 1972-6325
https://www.tpmap.org/

Open Access

COMPLEX MANAGEMENT OF WORSTER-DROUGHT SYNDROME: A CASE REPORT OF DOUBLE HEMIPLEGIA AND PSEUDOBULBAR PALSY IN A 13-YEAR-OLD

SHALINI PRIYA NANDAGOPAL[1], KOUSALYA KUMAR[1], DHANASANGARI M[1], ELILARASI S[1], DR. U. DAMEL LAKSHMI[2]
[1]PAEDIATRICS, SAVEETHA MEDICAL COLLEGE AND HOSPITAL, SAVEETHA INSTITUTE OF MEDICAL AND TECHNICAL SCIENCES, SAVEETHA UNIVERSITY, CHENNAI, INDIA
[2]ASSOCIATE PROFESSOR, DEPARTMENT OF PHYSIOLOGY, SREE BALAJI DENTAL COLLEGE & HOSPITAL, CHENNAI, INDIA.
CORRESPONDING AUTHOR: DHANASANGARI M

ABSTRACT

Introduction: Worster-Drought syndrome (WDS) is a rare congenital pseudobulbar palsy causing speech and feeding difficulties due to impaired corticobulbar pathways. Symptoms include drooling, chewing difficulties, dysarthria, and motor impairments, often categorized under cerebral palsy.

Case Report: We report a 13-year-old female with cerebral palsy (double hemiplegia), pseudobulbar palsy, global developmental delay, and speech delay. Key features included drooling since birth, chewing difficulty, and inability to speak. Birth history revealed severe asphyxia and meconium aspiration, requiring a 20-day NICU stay. Developmental delays were evident, with head control at one year and ambulation at two years. Examination showed left-predominant spasticity, hyperreflexia, muscle wasting, dynamic Achilles tendon contracture, and right-sided facial asymmetry. MRI revealed peri-insular temporal gliosis. Multidisciplinary management with speech, oral motor, and swallowing therapy was initiated, emphasizing continued follow-up and home-based exercises.

Results: The case underscores the complexity of managing CP with pseudobulbar palsy, necessitating targeted therapy for motor, speech, and feeding impairments.

Conclusion: Early multidisciplinary intervention in WDS can significantly improve communication, feeding, and daily functioning, enhancing the quality of life.

INTRODUCTION

Worster-Drought syndrome (WDS) is a rare form of congenital pseudobulbar palsy, characterized by impaired coordination and weakness of the muscles responsible for speech, swallowing, and facial movements. It arises from developmental disruptions in the corticobulbar pathways, which control cranial nerve function. Classified as a subtype of cerebral palsy, WDS often manifests with speech and feeding difficulties, including drooling, chewing challenges, dysarthria, and delays in speech development. Motor impairments such as spasticity, hyperreflexia, and limb contractures are also common [1].

The aetiology of WDS is typically linked to perinatal brain injury, hypoxic-ischemic encephalopathy, or prenatal disruptions in corticobulbar pathway development. Neuroimaging studies, such as MRI, often reveal structural abnormalities, including gliosis, cortical dysplasia, or other white matter changes, particularly in areas associated with corticobulbar and corticospinal tracts [2].

The syndrome poses significant challenges to daily functioning due to its combined effects on communication, feeding, and motor abilities. Management requires a multidisciplinary approach, involving neurology, speech

therapy, occupational therapy, and other specialists. Early diagnosis and intervention are critical to optimizing outcomes and improving the quality of life for affected individuals and their families [3]. The following case highlights the complexity of managing WDS, with a focus on its clinical presentation, diagnostic findings, and multidisciplinary management.

CASE REPORT

This report describes a 13-year-old female with a diagnosis of cerebral palsy (CP), specifically double hemiplegia, associated with pseudobulbar palsy, global developmental delay (GDD), and predominant speech delay. Her medical history revealed significant birth complications, including severe asphyxia and meconium aspiration, leading to a prolonged 20-day stay in the neonatal intensive care unit (NICU). Her antenatal history was unremarkable, with no maternal infections, substance exposure, or significant prenatal concerns.

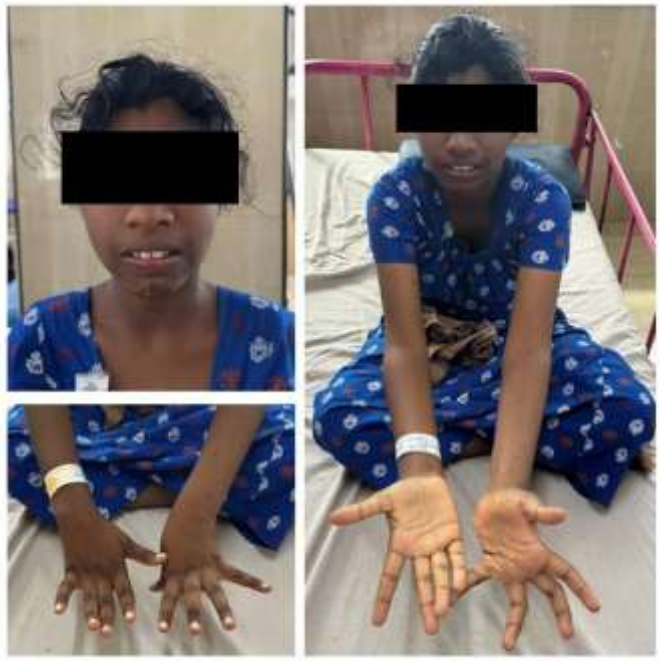

FIGURE 1- Profile of a 13-year-old with double hemiplegia and persistent drooling of saliva

The child's early development was markedly delayed. She achieved head control at one year of age and ambulated independently at two years. Fine motor skills showed a clear right-hand dominance, with severely limited function of the left hand. Speech development was notably impaired, and she remained non-verbal with persistent drooling and difficulty chewing. No significant history of seizures or other systemic illnesses was noted.

On examination, she exhibited spasticity in both upper and lower limbs, more pronounced on the left side. Muscle wasting was evident, particularly in the affected limbs. Reflex examination revealed hyperreflexia and dynamic contracture of the Achilles tendon bilaterally. Cranial nerve assessment showed Exaggerated jaw jerk reflex, Deviation of the mouth to the right. Absent gag reflex. Limited tongue movements, suggestive of significant pseudobulbar involvement. The child's anthropometric measurements remained within the 25th to 50th percentile despite significant swallowing difficulties. An IQ assessment performed at the Child Developmental Centre revealed severe intellectual disability, with an IQ score below 20.

Magnetic resonance imaging (MRI) of the brain revealed peri-insular temporal gliosis, a finding consistent with perinatal hypoxic-ischemic injury and reflective of damage to corticobulbar and corticospinal pathways.

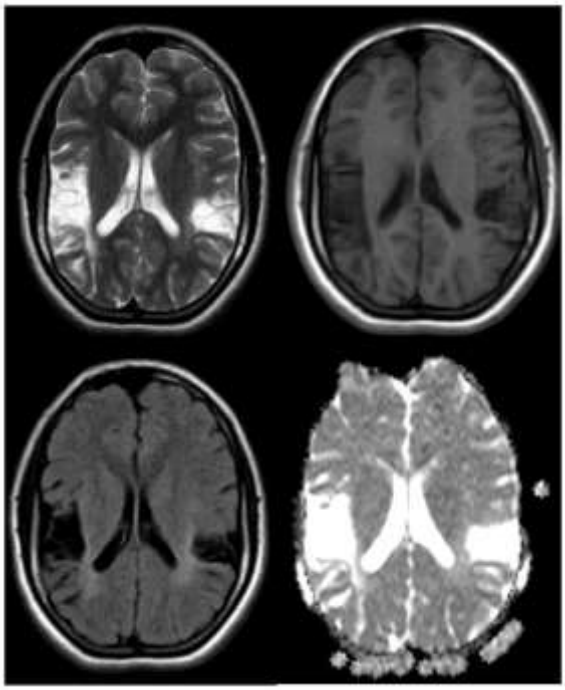

FIGURE 2- MRI Brain revealed peri insular temporal gliosis

The child received comprehensive care involving multiple specialties. Neurology management included muscle relaxants and physical therapy for spasticity control. Speech and language therapy was initiated to improve oral motor control, swallowing, and non-verbal communication skills. Otolaryngology addressed recurrent drooling and feeding challenges, with recommendations for techniques to improve swallowing efficiency. Ophthalmology assessed for any associated visual impairments. A home-based exercise program was designed to supplement professional therapy sessions. The importance of consistent follow-up was emphasized to monitor progress and adjust therapeutic interventions as needed.

DISCUSSION

Worster-Drought Syndrome (WDS) is a rare neurodevelopmental disorder resulting from abnormalities in the perisylvian region of the brain, which governs oral and pharyngeal motor functions. These abnormalities, often visible as bilateral perisylvian polymicrogyria on neuroimaging in about 15% of cases, are believed to arise between 12 and 16 weeks of gestation. The condition is attributed to impaired vascular supply or genetic factors, with familial cases reported in 15% of individuals regardless of imaging findings [4]. Neonatal feeding difficulties requiring specialized care are a common early manifestation, though WDS is not associated with perinatal injury.

The estimated prevalence of WDS is 3 in 100,000 live births, but the condition is likely underdiagnosed due to delayed recognition. Clinically, WDS presents with congenital pseudobulbar paresis, impairing the movements of the tongue, lips, jaw, and palate, leading to difficulties with feeding, speech, and swallowing. Additional features include motor delays, mild to moderate learning difficulties, behavioural challenges, epilepsy, and skeletal anomalies such as club feet [5]. Secondary complications, including gastroesophageal reflux, recurrent chest infections, and glue ear, further contribute to morbidity. Diagnosis is often delayed until after four years of age due to overlapping early symptoms with other neurodevelopmental disorders.

The coexistence of WDS with cerebral palsy poses additional challenges, particularly in cases of double hemiplegia. Bilateral motor impairments significantly limit mobility, posture, and balance, resulting in severe functional dependence. Secondary complications, such as joint deformities, contractures, and muscle weakness, exacerbate these difficulties, further reducing the child's quality of life [4,5].

Feeding and Swallowing Difficulties Feeding and swallowing difficulties are hallmark complications in children with WDS, particularly when associated with double hemiplegia. These challenges arise primarily due to

pseudobulbar palsy, which impairs motor control necessary for coordinated swallowing and feeding. In double hemiplegia, self-feeding, positioning, and food intake management become profoundly difficult [6]. Dysphagia, poor bolus control, and aspiration risk, compounded by pseudobulbar palsy, often lead to aspiration pneumonia and malnutrition [7]. Gastroesophageal reflux is prevalent, further increasing aspiration risk and causing discomfort. Approximately 10% of affected children require gastrostomy feeding to ensure safe and adequate nutrition.

Speech and Oral Motor Impairments Speech and oral motor impairments are significant in WDS. Dysarthria, characterized by slurred or difficult speech, impedes effective communication, leading to frustration and social isolation [8]. Impassive facial expressions or difficulties with voluntary facial movements due to impaired suprabulbar control may exacerbate these challenges, despite normal emotional expression. Drooling, caused by impaired oral motor control, is a persistent issue, contributing to social stigma and skin irritation [8]. Dysfunctional oral muscles may also lead to jaw contractures, misaligned teeth, and unusual palate shapes, complicating eating, oral hygiene, and increasing the risk of tooth decay, particularly in tube-fed children.

Multidisciplinary Management The management of WDS with double hemiplegia necessitates a multidisciplinary approach to address motor and non-motor complications:

- **Physical Therapy**: Early intervention promotes mobility, maintains joint flexibility, and strengthens muscles, improving functional independence [9]. Key interventions include spasticity management through stretching and positioning, gait training with assistive devices, postural control exercises, and oro-motor therapy in collaboration with speech therapists [10]. Functional training focuses on enhancing activities of daily living (ADLs) and upper limb coordination [11]. Respiratory physiotherapy may be needed for aspiration risks.

- **Speech and Language Therapy**: Oral motor exercises enhance swallowing functions and prevent aspiration. Augmentative and alternative communication (AAC) systems, including gestures, sign language, and communication aids, are essential for children with severe speech impairments [4].

- **Nutritional Support**: Monitoring hydration and caloric intake is critical to prevent malnutrition and dehydration, especially in children with dysphagia. Tailored feeding strategies and caregiver education on safe swallowing techniques are essential [12]. For severe dysphagia, percutaneous endoscopic gastrostomy (PEG) tubes may be required [7].

- **Pharmacological and Surgical Interventions**: Medications, such as anticholinergics for drooling and muscle relaxants for spasticity, play a role in symptom management. Surgical options, including tendon lengthening, baclofen pumps, or botulinum toxin injections, may improve spasticity and reduce drooling [13].

- **Behavioural and Cognitive Therapies**: Addressing intellectual impairments and behavioural challenges through specialized educational support ensures better developmental outcomes.

Anthropometric and Developmental Monitoring Despite feeding difficulties, children with tailored feeding strategies often maintain anthropometric measurements within normal percentiles, reflecting the effectiveness of these interventions. However, cognitive and educational support is critical for children with intellectual disabilities, ensuring optimized developmental outcomes.

Differential diagnosis includes conditions with overlapping motor and bulbar dysfunction. Cerebral palsy (spastic quadriplegia) and bilateral perisylvian polymicrogyria are key considerations due to shared pseudobulbar features, confirmed by MRI findings. Ischemic causes, such as perinatal stroke, arterial ischemic stroke, or Moyamoya disease, can result in progressive spasticity and motor deficits. [14] Metabolic disorders like mitochondrial diseases, leukodystrophies (e.g., metachromatic leukodystrophy, adrenoleukodystrophy), and genetic syndromes should be considered, particularly with systemic or progressive features. Structural brain abnormalities (e.g., corpus callosum dysgenesis, Chiari malformation), post-infectious encephalitic sequelae, and congenital TORCH infections further expand the differential, highlighting the need for neuroimaging, genetic testing, and metabolic workup for accurate diagnosis.

CONCLUSION

This case underscores the complexities of managing Worster-Drought Syndrome (WDS), particularly in the presence of double hemiplegia, where significant challenges in motor, speech, and feeding functions are evident. Early diagnosis and timely, targeted interventions, especially in oral motor and speech therapy, are critical for improving functional outcomes. A comprehensive, individualized care plan involving a multidisciplinary team—including neurologists, physiatrists, speech-language pathologists, and occupational therapists—is essential to address the diverse needs of affected children and mitigate complications [2].

Sustained therapeutic interventions not only enhance communication and feeding skills but also improve the overall quality of life for children with WDS and their families. Increased awareness among healthcare professionals and further research into long-term outcomes and optimal management strategies are vital to improving care and prognosis for individuals with WDS.

REFERENCES

1. Clark M, Carr L, Reilly S, Neville BG. Worster-Drought syndrome, a mild tetraplegic perisylvian cerebral palsy: review of 47 cases. *Brain*. 2000;123(Pt 10):2160-70. doi:10.1093/brain/123.10.2160.
2. Coulthard E, Hutchinson M, Hall M. Worster-Drought syndrome: clinical features, diagnosis, and management. *Dev Med Child Neurol*. 2008;50(5):377-82.
3. Boyle L, Johnson A, Green K. The role of multidisciplinary care in managing Worster-Drought syndrome. *J Pediatr Neurol*. 2011;9(3):267-73.
4. Simons J, et al. Management of hemiplegia in children: a multidisciplinary perspective. *J Pediatr Neurol*. 2018;45(5):321-7.
5. Jones K, et al. Neurodevelopmental disorders in pediatrics: a multidisciplinary approach to management. *J Pediatr Rehabil Med*. 2017;10(2):85-92.
6. Simons J, et al. Approaches to the management of feeding and swallowing disorders in neurologically impaired children. *J Pediatr Neurol*. 2017;44(3):209-17.
7. Meara JG, et al. Multidisciplinary approach to feeding difficulties in children with cerebral palsy and related conditions. *J Clin Pediatr Gastroenterol*. 2019;33(5):345-52.
8. Meara JG, et al. Worster-Drought syndrome: a review of pathophysiology and management. *J Clin Neurosci*. 2019;65:21-30.
9. Sheehan A, et al. Early intervention and its impact on functional outcomes in children with cerebral palsy and related neurodevelopmental disorders. *Dev Med Child Neurol*. 2016;58(4):307-12.
10. Chitrada R, Kamalakannan M, Natarajan S, Anbu V, Venkat A, Shathurshini B, et al. Assessing the efficacy of intermittent intensive physical therapy in pediatric cerebral palsy. *INTI Journal*. 2024. Available from: https://iuojs.intimal.edu.my/index.php/intijournal/article/view/466.
11. A study on the effectiveness of task-oriented strength training to enhance upper limb motor function in children with cerebral palsy. *Indian J Physiother Occup Ther - Int J*. 2024;18(5):376-80. doi:10.37506/z230pd05.
12. Jones K, et al. Swallowing and feeding disorders in cerebral palsy: pathophysiology and management. *J Pediatr Rehabil Med*. 2018;10(2):91-8.
13. Sheehan A, et al. Feeding and swallowing difficulties in children with neurodevelopmental disorders. *Dev Disabil Res Rev*. 2016;18(4):1-10.
14. Dhanush Balaji S, Rangan AS, Prasanna Karthik S, Gopalakrishnan KV, Balaji S, Manoj Kumar S. Study to estimate the NIHS score and volume of stroke in predicting 30-day mortality in acute ischemic stroke patients. *Ro Med J*. 2024;71(2):105–9. doi:10.37897/RMJ.2024.2.6.