Worster-Drought syndrome: poorly recognized despite severe and persistent difficulties with feeding and speech

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LIST OF ABBREVIATIONS

CBPP	Congenital bilateral perisylvian polymicrogyria
SOMA	Schedule for Oral–Motor Assessment
WDS	Worster-Drought syndrome

AIM Worster-Drought syndrome (WDS), or congenital suprabulbar paresis, is a permanent movement disorder of the bulbar muscles causing persistent difficulties with swallowing, feeding, speech, and saliva control owing to a non-progressive disturbance in early brain development. As such, it falls within the cerebral palsies. The aim of this study was to describe the physical and neuropsychological profiles of children with WDS.

METHOD Forty-two children with WDS (26 males, 16 females; mean age 7y 10mo, SD 3y 1mo; range 2y 6mo to 16y 5mo) were studied prospectively using a standard protocol.

RESULTS All of the children had severe bulbar dysfunction; 36 out of 42 had feeding difficulties and 23 of 38 had unintelligible speech, which was poorly compensated for by augmentative communication. There were accompanying disturbances in cognition (mean non-verbal IQ 59), behaviour (12/40 attention-deficit–hyperactivity disorder [ADHD]), social communication (8/42 autism), and epilepsy (12/39). The severity of bulbar dysfunction and impact of additional impairments made it difficult to use formal assessments.

INTERPRETATION WDS causes severe and persistent bulbar dysfunction that is often accompanied by additional impairments, as in other cerebral palsies. Speech prognosis is particularly poor. Early diagnosis with appreciation of the underlying neurology would encourage critical evaluation of interventions and long-term planning to improve outcome.

WDS was originally defined as congenital suprabulbar paresis,[1] without specifying the level of lesion. However, the term has been used for acquired lesions,[2] functional epileptogenic variants,[2] and even isolated dysarthria.[3] There are only a few retrospective case series, if the term is confined to congenital spastic paresis (great difficulty using bulbar muscles with no wasting or fasciculation; exaggerated jaw jerk) causing permanent bulbar dysfunction, resulting in disorders of saliva control, swallowing, airway protection, feeding, and speech.[1,4,5] The underlying abnormality is thought to be in the perisylvian cortex (congenital bilateral perisylvian polymicrogyria [CBPP] occurs in around 15/100 of affected individuals[5]) and may be a continuum of bilateral perisylvian syndromes.[6,7] A genetic aetiology appears to be important in about 15/100[8] of cases and there is evidence of a vascular aetiology in some individuals.[3,8]

The differential diagnosis is relatively straightforward. There must be difficulty from infancy (although exact expression varies with severity and developmental stage), which excludes acquired and progressive conditions. The main motor involvement is bulbar, distinguishing it from suprabulbar palsy as part of another diagnosis, such as severe spastic tetraplegia. Cases in which bulbar involvement is lower motor neuron, or due to motor unit or muscle disorder, are easily excluded as the tongue is weak, wasted, and tremulous, and jaw jerk is absent. Structural causes must be considered. Tongue tie is simple to eliminate as a cause of reduced tongue movement as it does not affect the lips, jaw, or palate. However, structural causes of velopharyngeal insufficiency (e.g. occult submucous cleft) can cause confusion, as some children compensate for the loss of air through the nose by articulating at the vocal folds rather than with the tongue or lips. However again, careful examination would show that oromotor movements are intact.

Another important differential diagnosis is childhood apraxia of speech or developmental verbal dyspraxia in which the precision and consistency of movements underlying speech are impaired. This leads to inconsistent error production, impaired coarticulatory transitions between sounds and syllables, and inappropriate prosody.[9] There is no weakness on formal examination and no difficulty with involuntary motor control of chewing, or swallowing, etc. However, many children given this diagnosis also have 'messy eating' or mild dribbling.[9] This can cause confusion as the pyramidal motor speech problems in WDS may be manifest as symptomatic dyspraxic or dysarthric speech, and the distinction from

developmental verbal dyspraxia rests on the severity and multimodality of the bulbar involvement, confirmed by signs of suprabulbar paresis on examination.

WDS meets the international criteria for cerebral palsy (CP), being a 'permanent, but not unchanging disorder of movement and of motor function due to a non-progressive interference, lesion or abnormality of the developing/immature brain'. Affected children commonly have a mild tetraplegia causing late motor milestones, although they do walk.[5] As with other types of CP, there are often additional impairments in behaviour and learning, and epilepsy may be present.[5] A CP study[10] identified WDS in 0.9/100 of participants, suggesting a prevalence of 2 to 3 per 100 000 live births. From our experience this is probably an underestimate, as late diagnosis is common (mean age 5y in this study), many CP registers use 5 years as the optimal ascertainment age,[11] and most do not recognize WDS. Furthermore, many children are not diagnosed but managed entirely on the basis of their symptoms. As a consequence, systems for assessing and managing this symptom complex are mostly not available.

METHOD

Participants

Thirty-nine children older than 4 years with congenital suprabulbar paresis were consecutively recruited over 18 months at Great Ormond Street Hospital from clinics for augmentative communication, neurodevelopment, dysphagia, and neurology. Four years was chosen as the minimum entry age because motor findings can be transient in younger children (any motor speech problem should be evident by this age), and to improve compliance with assessment. In addition, three younger siblings (2y 6mo, 2y 8mo, 3y 5mo) with a secure diagnosis were enrolled. In total, 42 children with congenital suprabulbar paresis (26 males, 16 females; mean age 7y 10mo; SD 3y 1mo; age range 2y 6mo to 16y 5mo) were studied prospectively.

This study was approved by the Research Ethics Committee for Great Ormond Street Hospital and the Institute of Child Health. Informed consent was received from all parents or guardians before assessment. Anonymized data were used throughout the analysis.

Measures

Participants followed a standard protocol (Table SI, supplementary material published online) with data collected by four speech and language therapists, two paediatric neurologists, and a radiologist. Feeding (28 children) and swallowing (11 children) studies were restricted to those less severely affected, and contraindicated when clinical assessment suggested that they may be unsafe. Reliability was monitored by independent evaluation of video-recorded assessments, and discrepancies were resolved by consensus.

Statistics

Descriptive statistics were used. In the case of data being unavailable for any individual, that individual was removed from analysis of the variable, and the denominator therefore reflected the number of valid data points. χ^2 tests were used to explore relationships between categorical variables; *t*-tests were used to compare group data.

RESULTS

General

The results are shown in Table I. Children presented early (mean 7mo) but were diagnosed late (mean 5y). A family history of WDS (4/36), speech delay (5/36), or seizures (9/36) was common. Twelve out of 39 participants had epilepsy, often with multiple seizures (5/12) and a history of convulsive status (5/12). Twelve out of 40 participants had a diagnosis of ADHD and 8/39 had a diagnosis of autism made by local

Table I: Cross-sectional clinical data

Characteristic		*n*/Total
Sex, M/F		26/16
Mean (SD) age		
Presentation	7mo (10mo)	
Diagnosis	5y (2y 10mo)	
Assessment	7y 10mo (3y 1mo)	
Initial presentation		
Feeding		22/42
Developmental concern		10/42
Language delay		9/42
Seizures		1/42
Family history		
WDS		10 Children, four families
Speech delay (not WDS)		5/36 Families
Seizures		9/36
Motor milestones		
Mean (SD) age sat	11mo (4mo)	
Mean age walked	1y 11mo (9mo)	
Epilepsy		
Frequency		12/39
Mean (SD) age of first seizure	2y 7mo (1y 9mo)	
Neuropsychiatric disorder		
ADHD		12/40
Autism spectrum disorder		8/39
Cognitive assessment, mean (95% CI)		
Non-verbal IQ	59 (50–68)	
Receptive DQ[a]	62 (52–72)	
Expressive DQ (spoken content)	40 (35–45)	
Examination		
High arched palate		5/29
Jaw contractures		10/42
Limb contractures		5/41
Abnormal eye movements		5/42
Pyramidal signs		38/40
Poor imitation of gestures[13]		15/23
<4th centile for upper limb speed/dexterity[14]		27/28
Investigations		
Perisylvian polymicrogyria		5/37
Abnormal electroencephalography		13/32
Neurogenic bulbar electromyography		5/13

[a]Developmental quotient= (age equivalent at assessment/chronological age at assessment) ×100. WDS, Worster-Drought syndrome; ADHD, attention-deficit–hyperactivity disorder; CI, confidence interval; DQ, developmental quotient.

child development teams using ICD-10 criteria. This was congruent with behavioural screening (Strengths and Difficulties Questionnaire),[12] which showed significant differences between the WDS group and normal population across all five behaviour scales (independent samples *t*-test, $p<0.001$), particularly striking for hyperactivity and peer relationships (Figure SI).

Thirty-eight out of 40 participants had mild pyramidal limb signs (brisk deep tendon reflexes with mild increase in tone but without obvious weakness), with 15 out of 23 in the dyspraxic range for imitation of gestures[13] and 27 out of 28 below the 4th centile for rapid and precise upper limb movements.[14] These factors could all affect signing and other augmentative communication methods.

Oromotor examination, drooling, and feeding

All children had multiple abnormalities on oromotor examination of the lips, tongue and jaw (restricted jaw opening precluded systematic observation of the palate) (Table II). Eleven out of 20 children had dissociation of facial expressions (e.g. could not smile on request or through imitation but produced a reflex smile in response to emotion), and 10 of 38 had a jaw contracture.

Eleven children had required neonatal care for swallowing problems and compromise to their airway, including one child who subsequently underwent an epiglottopexy. All 42 children drooled beyond 2 years. This improved and become intermittent with age, often returning whenever the child had to concentrate (e.g. on school work). Three children had undergone submandibular duct transposition with limited success, despite repeat operations in one individual. Twenty-three out of 42 children had tried but discontinued medication because of unpleasant side-effects and supply issues. Only six children remained on medication, and then only intermittently on special occasions. Thirty-two out of 41 participants drooled during assessment.

Families tended to underreport feeding issues, but detailed questioning (Table III) revealed that 29 out of 41 participants had difficulty establishing milk feeds as infants (compared with 1/10 to 15/100 of infants in the general population[15]), 40 out of 42 participants experienced problems during weaning, and 10 of 42 required prolonged tube feeding. Most reported managing more complex food over time. At the time of assessment, 5 of 42 participants were dependent on tube feeds, 31 out of 42 needed dietary modification, and only 6 of 42 managed a full adult diet. However, many children who required dietary modification perceived themselves as 'managing a careful normal diet' with the caveat that they avoided certain textures (e.g. apple skins, meat, crisps), chose 'soft foods', and chopped or mashed it well. Many reported oral hypersensitivity to certain textures and also to tooth brushing, defined as causing apparent severe discomfort.

A structured feeding assessment (Schedule for Oral–Motor Assessment; [SOMA][16]) was attempted in 28 children. Refusal rates were high, ranging from 5 out of 28 for liquid to 11 of 28 for purée. Three children completely refused to undertake the assessment; nine completed the assessment with normal results, and in 11 assessment was incomplete, with normal results where tested. Only five children had abnormal scores (three for multiple textures), despite the fact that 21 of the 25 children tested needed dietary modification at the time. There was no clear pattern to the abnormalities seen. Thus, SOMA findings did not reflect the reported feeding difficulties.

Informal feeding observation in these 28 children revealed functional difficulties and behavioural adaptations that were not scorable on the SOMA. Children compensated for reduced lip movement by tilting their chin upwards to reduce anterior spill of saliva and food, and by using their hands to achieve lip seal. Similarly, they overcame impaired tongue movements by using their fingers to place food between the teeth or by not chewing food but instead mashing it against the hard palate. Many overfilled the mouth, perhaps compensating for food falling out or because of reduced oral awareness. Swallowing was often initiated by tipping the head back to use gravity or by placing a supporting hand on their chin. Food residues commonly stuck to the hard palate or cheeks and had to be cleared manually after meals otherwise residues

Table II: Oromotor neurological assessment

Oromotor abnormality		*n*/Total
Open mouth at rest		38/39
Drooling		32/41
Dissociation of facial expression		11/20
Jaw contracture[a]		10/38
Jaw jerk		36/40
	Impaired	Absent
Movement	*n*/Total	*n*/Total
Vertical jaw	11/34	6/34
Lateral jaw	5/30	22/30
Tongue protrusion	20/37	5/37
Lateral tongue	17/37	18/37
Tongue elevation/depression	13/34	18/34
Lip (closure/spread/round/smacking)	27/36	6/36

[a]One child had ankylosis requiring surgery.

Table III: History of feeding and associated difficulties

Feeding characteristics	*n*/Total
History of early feeding difficulties	
Suckling	29/41
Weaning puree	34/41
Weaning lumps	37/42
Weaning mature diet	40/42
Prolonged tube feeds	10/42
History	
Gastro-oesophageal reflux	22/41 (7 surgery)
Recurrent chest infection	25/38
Recurrent otitis media	26/41 (22 grommets)
Current feeding	
Normal diet	6/42
Soft/careful diet	16/42
Purée/chopped diet	15/42
Tube feeds	5/42
Current drooling problems	39/41

could remain in the mouth, even overnight. Children used their hands or napkins to mask their mouths during eating so that superficially eating appeared acceptable, and overall they tolerated significant feeding abnormality.

Videofluoroscopy was attempted in 11 children. (Table SII) Two children refused outright and many refused certain textures, perhaps reflecting a learned aversion by children with feeding difficulties. All nine participants had abnormal assessments, with seven having multiple abnormalities and two not completing the assessment. The children cooperated best for purée (3/11 refused) and also had fewest abnormalities for this texture. Liquids were less acceptable (4/11 refused) and appeared generally to be more difficult to handle (perhaps as the more rapid transit unmasked the coordination difficulties). Solids were refused by 8 of 11 children, making results for this texture difficult to interpret.

Speech, language, and cognition

Many of the children had learning difficulties, and on a paired-samples *t*-test there was no significant difference between non-verbal IQ scores (mean=59, confidence interval [CI] 50–68) and receptive developmental quotient (DQ) scores (mean=62, CI 52–72). Expressive DQ was extremely difficult to measure owing to intelligibility issues. Preliminary analysis gave a mean expressive DQ of 40 (CI 35–45) and a significant difference between receptive and expressive DQ scores on a paired-samples *t*-test (t[17]=2.57, p<0.02, the η^2 statistic [0.28] indicating a large effect size). To examine whether this simply reflected intelligibility rather than true difference in expressive intent, nine children across the full receptive DQ range were assessed by the information-carrying content of their communication, including responses through augmentative communication (Fig. 1). Augmentative communication allowed less cognitively able children to achieve expressive scores similar to their receptive abilities, suggesting that the observed receptive–expressive gap was largely a reflection of articulatory difficulties that could be overcome with appropriate support. However, in children with DQ in the normal range, a large receptive–expressive gap remained, implying that these children had not achieved their potential with current provision and use of augmentative communication.

By report, many children had received significant direct speech therapy without developing functional speech. However, there were a few children who did acquire some expressive language very late (e.g. four children had no words at 3–4y but had 15–20 unclear words by 6–8y). Thirty-two of 42 children relied on augmentative communication systems (Table SIII).

Twenty-three children out of 38 were unintelligible at single word level,[17] even to their familiar listeners (Table IV). Twenty-four of 37 had a severe dysarthric pattern causing difficulty with articulatory placement, especially tongue tip sounds, hypernasality, and gross nasal escape. They often produced predominantly velar sounds (k, g) made with the back of the tongue against the soft palate. However, 5 out of 37 participants made more typical dyspraxic-type errors, with vowel distortion, different targets produced on consecutive attempts, and 'groping' for articulatory placement, including one child with bilateral perisylvian polymicrogyria and a family with dysarthric and dyspraxic members. The remaining 8 of 37 children could not be categorized and often displayed mixed features. Some children had difficulty with voice production, with use of lower register ('creaky') voice further impeding intelligibility.

Communication was often additionally compromised by poor social communication skills, including difficulties with initiating conversation, maintaining a topic chosen by adults, and failing to notice change of topic. However, there was no link between autism and intelligibility. Independent-samples *t*-tests were conducted to compare the non-verbal IQ scores for children with autism (mean 50) and those without autism (mean 61), for children with ADHD (mean 56) and those without ADHD (mean 60), and, finally, for those with epilepsy (mean 54) and without epilepsy (mean 62); there were no statistically significant differences.

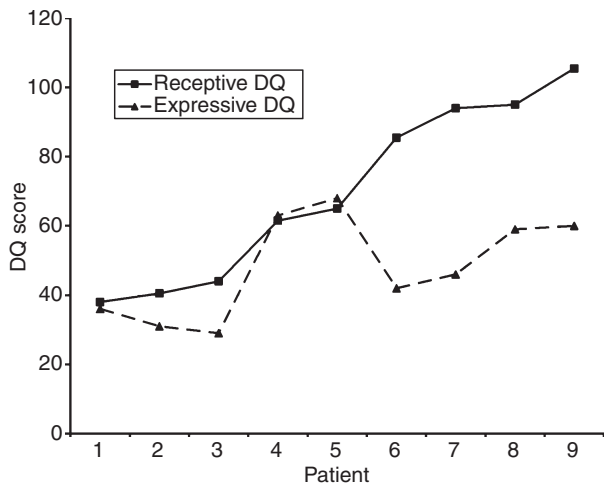

Figure 1: Expressive developmental quotient (DQ; including multimodal communication responses) of nine children compared with their receptive DQ. DQ= (age equivalent at assessment/chronological age at assessment) ×100.

Table IV: Intelligibility findings for children with Worster-Drought syndrome

Level[17]		Frequency
Normal intelligibility	Speech intelligible, not affected by motor disorder	0/38
Mildly unintelligible	Many single words/ phrases in/out context	9/38
Moderately unintelligible	Many single words, short phrases in context	6/38
Severely unintelligible	Some single words	10/38
Profoundly unintelligible	Open vowels only, unintelligible to familiar adults without contextual clues	13/38

Other investigations

Five out of 37 children had CBPP. In 13 out of 32 children electroencephalography was abnormal, often showing spike–wave complexes over the Sylvian regions.

In 5 out of 13 children (two with CBPP), a tongue electromyogram (EMG) was abnormal, showing neurogenic changes suggestive of lower motor neuron involvement, which has been reported before in CBPP.[18] There was no significant difference between the group with neurogenic changes and those in whom the EMG was normal in terms of oromotor examination, intelligibility, and current diet, but the neurogenic group had higher mean non-verbal IQ (90 vs 53) and mean receptive DQ (71 vs 47) scores. This might have suggested a non-cortical lesion, as found by other groups,[19] except that two children (both with non-verbal IQ>75) had CBPP. pH studies carried out in four children showed significant reflux.

DISCUSSION

Oromotor examination, drooling, and feeding

Oromotor impairments (lip, jaw, tongue) appeared to be independent, with the tongue most frequently and severely affected. Severe tongue and lip impairments commonly accompanied difficulties with feeding, intelligibility, reflux, and aspiration, although not reaching statistical significance. Severely restricted jaw movement was associated with a history of aspiration (χ^2, p=0.042), and during feeding assessment these children needed help to stabilize their jaw to allow safe feeding. However, there was no link between degree of oromotor examination abnormality and receptive language, non-verbal IQ, neuropsychological profile, sex, or presence of perisylvian polymicrogyria.

Thirty-nine children out of 41 (39/41) had significant drooling, which has previously been correlated with reduced swallowing and oromotor control.[20] Where performed, surgery appeared unhelpful. Twenty-six out of 32 children chose not to take medication and used behavioural techniques such as chin posture, wiping the mouth, and conscious swallows to reduce social impact.

Forty out of 42 participants had problems with suckling and weaning, but were often not diagnosed until school age. Those with difficulty from birth were significantly more likely to also have a history of gastro-oesophageal reflux (χ^2, p=0.01) and aspiration (χ^2, p=0.04). Children and parents become habituated to feeding difficulties so that history-taking had to ask specifically about feeding milestones, dietary modification, and eating behaviour, and corroborate this with direct clinical observations of feeding using the child's preferred foods. Reported oral hypersensitivity appeared to be a behavioural rejection of certain stimuli based on previous unpleasant experiences (including gastro-oesophageal reflux and constipation), rather than a neurological sensory issue. In some children it further compromised feeding and made dental and oral hygiene extremely difficult.

Direct feeding observation gave better information than formal assessment, and showed the children using many compensatory strategies to mask difficulties and consciously choosing less challenging food. The SOMA had high refusal rates (probably because the children knew their limitations) and did not capture the difficulties or detect compensatory movements, perhaps because the SOMA was devised to assess oromotor skills during weaning of younger children, and in WDS the feeding pattern is severely disordered rather than delayed. Videofluoroscopy was restricted to less severely affected children to minimize aspiration. Even so, refusal rates were extremely high, perhaps because these children were mobile and hyperactive, and had strong food aversion. Despite this, videofluoroscopy gave important information on swallowing that was not available from direct observation.

Communication

These children experienced multiple barriers to communication, including motor speech problems, social communication difficulties, hyperactivity, and conductive hearing loss. In 23 out of 38 children, motor speech was so severely affected that the children were effectively unintelligible, a significant, practical barrier to accessing learning and participating in friendship groups. Thirty-two out of 42 children used alternative communication systems, but they often did not achieve their communicative potential, with large discrepancies between language understanding and expression, particularly in the case of more able children. This communication gap may have resulted from difficulties in accessing equipment appropriately matched to the child's profile[21] or in obtaining adequate support for effective equipment use, as well as from the effects of other aspects of the child's profile (e.g. hyperactivity, dyspraxia).

The observed severity and longevity of intelligibility difficulties mean that all direct speech work should be carefully evaluated for improvements in functional intelligibility. It may be best to assume that augmentative and alternative channels of communication will be required and to introduce these early so that children have the 'tools' to develop expressive communication.[22] It is difficult for this group to achieve successful use of alternative channels of communication because of their reduced fine motor, attention, and social communication skills and their normal mobility; therefore, devices need to be robust and portable, and trial of loan equipment may be necessary.

Methodological problems

Completion of assessments was difficult for many reasons (food aversion and inability to understand instructions, to comply with adult direction, to sustain attention, or to repeat items for scoring purposes), although none reached significance, perhaps because of small numbers or because most of the group had some difficulty in these areas. Assessment by experienced clinicians using real situations appeared to be the most successful solution.

CONCLUSION

Children with WDS commonly present to community services with feeding problems and speech delay. Early diagnosis of this neurological condition and its identification as distinct from a maturation or developmental disorder is important because oromotor difficulties are likely to persist and do not appear to respond to conventional therapies, and affected chil-

dren may have multiple additional impairments that require appropriate multidisciplinary support to optimize outcome.[23]

Feeding problems are extremely important, leading to poor growth and developmental outcomes,[24] decreased motivation, increased irritability, serious pulmonary consequences,[25] and stressful mealtimes. Professionals actively need to ask about specific feeding behaviours and milestones as families often become habituated to the abnormal situation. There is a high incidence of non-compliance with formal assessment, so real-time observation of feeding is invaluable. The feeding difficulties are complex and may include gastro-oesophageal reflux, aspiration, constipation, dehydration, malnutrition, and secondary behavioural components, as well as difficulties with drooling, dental hygiene, and oral hypersensory issues.

The prognosis for speech development for children with WDS is poor, and there is always a marked discrepancy between receptive and expressive language skills. Both dysarthric and dyspraxic elements contribute to their intelligibility difficulties, and, although they are reported not to respond to direct speech work, interventions shown to be useful for children with CP and dyspraxia[26] could be considered with careful evaluation of results. Early, appropriate, and well-supported alternative communication programmes that cater for ADHD, autism spectrum, and motor impairments would be important to optimize communicative outcome. Outcome measures would need to include all communication functions and evaluate integrated use of speech, signing, facial expression, and equipment.

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SUPPORTING INFORMATION

Additional supporting information may be found in the online version of this article:

Figure S1: Strength and difficulties questionnaire results for children with Worster-Drought syndrome compared with the normal population.

Table SI: Assessment protocol

Table SII: Findings on videofluoroscopy

Table SIII: Communication methods used by children with Worster-Drought syndrome

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