Worster-Drought syndrome, a mild tetraplegic perisylvian cerebral palsy

Review of 47 cases By Maria Clark, Lucinda Carr, Sheena Reilly* and Brian G. R. Neville

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Extract:
A retrospective case-note analysis was undertaken of 47 children with a congenital upper motor neurone bulbar palsy (excluding pure speech dyspraxia) to clarify the phenotype of Worster-Drought syndrome (WDS) and to record its associated features and complications. The results revealed that the study children had significant bulbar problems (with 80% still needing a modified diet and a similar number using augmentative communication methods at last review). There were also high rates of predictable bulbar complications (86% had dribbling, 60% had glue ear, gastroesophageal reflux in 40%, history of poor nutrition in 40% and aspiration in 40%). Most of the children had additional complex impairments (91% had mild pyramidal tetraplegia, 81% learning difficulties, 60% congenital defects, 41% neuropsychiatric problems and 28% epilepsy). Over half of the children had significant medical problems in the first year, but mean age at diagnosis was 6 years. There were no obvious causes in pregnancy or birth. Six children had a family history of WDS and 32% (12/37) had abnormal neuroimaging including five with bilateral perisylvian polymicrogyria. In our experience, WDS is not uncommon, is relatively easily diagnosed and is crucial not to miss as the management of these children’s multiple impairments is complex and requires a careful team approach. WDS falls clearly within the cerebral palsies as a syndrome that includes motor impairment arising from static damage to the brain in early life. The common presence of cognitive, behavioural and seizure impairments strongly supports the cerebral cortical (presumably perisylvian) localization. Its core elements are a suprabulbar paresis, a mild spastic tetraplegia and a significant excess of cognitive and behavioural impairments and epilepsy. The complete overlap in phenotype between WDS and the bilateral perisylvian syndrome leads us to propose that they are the same condition. WDS is startlingly absent from epidemiological studies of the cerebral palsies and rarely diagnosed, presumably because of lack of clinical awareness of the condition and lack of major gross motor impairments.

http://brain.oupjournals.org/cgi/content/abstract/123/10/2160

Brain, Vol. 123, No. 10, 2160-2170, October 2000
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Thomas’s Story

For as long as we can remember Thomas has been tip toeing. At the age of three, Thomas’ paediatrician decided it was time to do something in consultation with Mary our physio they decided to put Thomas into plaster for three weeks, it was a difficult time as I was very pregnant with our other son Alex.

So Thomas was put into plaster the three weeks came and went Thomas had the plasters removed, and the first thing he did was go straight up on his toes.

The next idea was to try special shoes and boots, Thomas was fitted for boots, he wore them, but he managed to walk on his toes inside the boot. Next night splints were tried, Thomas would not wear them at night, so he would have them on for 10-30 minutes during the day, he didn’t like wearing the splints.

Thomas would even damage the heel part of his shoes enabling him to get on toes.

The problem carried on, and he didn’t grow out of it.

At the age of 7 Thomas was referred to a new paediatric orthopaedic surgeon Mr Trevitt who had started working at Worcester. He had successfully used botulinum injections as a muscle relaxant on children in Bristol (they also use the technique before performing surgery such as cutting the tendons to see if the surgery is likely to work )

These injections were to be used on Thomas, he would have 4 injections in each leg into his tendon and then after a few days would be put into plaster for 4 weeks. Thomas had the injections under general anaesthetic and then went into plaster, four weeks came and went the plasters were taken of yes you guessed it he was able to walk on his toes, to be fair his whole posture had improved but this was only short term.

Back on his toes it was felt by Mr Trevitt and Mary that surgery was the only solution left. In June of this year Thomas had his tendons cut, a technique known as Hokes procedure. A small incision was made into the tendon on either side. Thomas had three incisions in each leg. He went straight into heavy plasters, which were kept on for a week, then removed for lighter plasters, which stayed on for 5 more weeks. After 5 weeks the plasters came off, success Thomas stomped and walked heavily around the house, flatfooted for the first week, his legs were painfully thin. Each week we saw some improvement in his walking though we were unable to do many exercises, as this would involve him being on his toes, so patience was needed.

He now does try to go up on his toes but his tendons are so lose he cannot stay up. Two months on, his tendons are now tightening up this is the critical time. He is mainly walking properly and his posture is wonderful.

And we keep watching, fingers crossed and heels flat.

Andrew and Karen Hinks.

Many Thanks to Alan & Anne

Taking the high road: Alan Burns and his wife Anne from Haltwhistle, travelled 819 miles in their 1948 vintage Leyland Beaver lorry on a charity trip through some of the most glorious scenery in Scotland en route from Lockerbie to the Isle of Skye. As well as enjoying the scenery, they raised £856.80p for the Worster-Drought Syndrome Support Group, and accepting the cheque from Alan is family friend Dean Garcia from Hexham, who suffers from the rare complaint.