Isolated bulbar palsy and dysphagia in children with respiratory symptoms

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ABBREVIATIONS

gEMG	Genioglossus electromyography
VFSS	Videofluoroscopic swallow study

Abstract

Oropharyngeal dysphagia can cause chronic aspiration leading to significant respiratory symptoms. When dysphagia is diagnosed, an underlying cause is sought. We present a case series of 15 children diagnosed aged 6 months to 5 years (mean 2y 5mo; 11 males, four females) over a 6-year period, who were found to have an isolated bulbar palsy on genioglossus electromyography, with no accompanying neurological or neurodevelopmental disorder. Eight children had dysphagia but a normal EMG. In those with isolated bulbar palsy, management included thickened fluids (n=13), cooled boiled water (n=1), and nasogastric tube feeding (n=1). Follow-up over 1 to 8 years (mean 5y) showed complete resolution in six children, improvement in four children, and no improvement in five children (including two requiring fluids via a gastrostomy). Eight children no longer had any respiratory symptoms. Isolated bulbar palsy is under-recognized and has not been reported previously as a cause of significant dysphagia in children.

A fully functioning neurological system is necessary for proper coordination of breathing and swallowing to protect the airway when drinking. Oropharyngeal dysphagia can result in inadequate airway protection during swallowing causing chronic pulmonary aspiration. Whilst the principal symptom of coughing/choking when drinking may be obvious, it may also be silent and the parents may be unaware of the issue. These children are often referred from primary or secondary care to respiratory paediatricians for assessment of symptoms such as acute life-threatening events in an infant, recurrent pneumonia, persistent wet cough, or intractable wheeze.[1] Sometimes the child is referred late with bronchiectasis already evident from years of aspiration damage. As part of the work-up of these respiratory issues, our speech and language therapist assesses for dysphagia.

There are multiple causes of dysphagia, including several neurological, neurodevelopmental, and anatomical issues, although in many children the reasons are unclear.[2] When appropriate we look for an underlying diagnosis and this may include a neurological assessment, even in typically developing children, as we wish to exclude bulbar palsy (affecting cranial nerves IX, X, XII motor nuclei in the medulla) using genioglossus electromyography (gEMG).[3] In one of the principal paediatric respiratory textbooks,[4] 'otherwise normal child with isolated aspiration' is listed in the long list of causes. Our experience shows some of those children have an isolated bulbar palsy with no other accompanying neurological or neurodevelopmental condition. We believe this is an under-recognized disorder that has not been reported and is not included in lists of causes in reviews of the subject. This case series highlights this diagnosis and looks at its management and prognosis.

CASE REPORTS

This was a retrospective electronic note review of children with dysphagia who presented to a tertiary respiratory service with chronic chest symptoms, and who were found to have an abnormal gEMG with an isolated bulbar palsy. We excluded those who had accompanying neurological or neurodevelopmental issues, including those with an identified pathology on magnetic resonance imaging (MRI) brain scan, epilepsy, and autism spectrum disorder. We included a group with dysphagia and a normal gEMG. The diagnosis of dysphagia was made by a speech and language therapist after clinical assessment and videofluoroscopic swallow study (VFSS). Depending on the findings on VFSS and other results, referral was made to a paediatric neurologist and neurophysiologist for further assessment to rule out neurological conditions, including bulbar palsy. Details of the assessments are in Appendix S1 (online supporting information).

The study was accepted as a service review/clinical audit by clinical audit committees at Royal Brompton (001819) and Chelsea & Westminster (LA427) Hospitals and formal ethical approval was deemed not necessary. All parents gave verbal telephone consent for us to include their children in the series.

RESULTS

Between December 2012 and October 2018 we identified 15 children with dysphagia causing respiratory symptoms who had an isolated bulbar palsy. Their mean age was 2 years 5 months (range 6mo–4y 11mo); 11 were male and four were female (Table 1). We also identified eight children with dysphagia who had a normal gEMG in whom we could not confirm isolated bulbar palsy. Their mean age was 4 years 3 months (range 1y 2mo–7y 11mo). Their details are given in Tables S1 and S2 (online supporting information).

Details are given here for those with isolated bulbar palsy. Presenting respiratory symptoms were persistent wet cough (*n*=7), wheeze (*n*=6), recurrent lower respiratory tract infections (*n*=3), and persistent dry cough (*n*=2); three children had multiple symptoms. Symptoms reported by the speech and language therapist clinical history were coughing when drinking (*n*=13), wet voice (*n*=1), and none (*n*=1); the initial assessment concluded silent aspiration suspected (*n*=8), no aspiration noted (*n*=5), overt aspiration (*n*=1), and refusal to drink (*n*=1). The age of first feeding symptoms was mostly 0 to 6 months (*n*=10), with some older (6–12mo *n*=2, >12mo *n*=2), and one with no reported symptoms.

Details of the first and last VFSS are in Table 2, alongside the latest clinical update. More details of the swallow characteristics are in Table S3 (online supporting information). The diagnostic VFSS resulted in children having thickened fluids at International Dysphagia Diet Standardisation Initiative[5] level 3 (*n*=12); level 2 (*n*=1); one child had cooled boiled water due to penetration of level 3; and one child started nasogastric feeding with tastes of breastmilk and purees by mouth.

What this paper adds

- Dysphagia may be due to an isolated bulbar palsy.
- Prognosis cannot be predicted although dysphagia resolved in one-third of patients.

All 15 children had an abnormal gEMG indicating bulbar palsy. Congenital myasthenic disorders were ruled out by appropriate EMG studies.[6] Fourteen children had a normal brain MRI scan, one child did not have a scan but was typically developing at 3 years 2 months (patient no. 4). All children had normal blood and urine tests for neurometabolic disorders, including negative acetylcholine receptor and antiMuSK antibodies, with normal acyl carnitine profiles; all had negative genetic testing for Fazio-Londe and Brown-Vialetto-Van Laere syndromes.

Nine children had a 24-hour pH study, which was abnormal in one child; one child with a normal study was treated for gastroesophageal reflux due to clinical symptoms. Seven children had a rigid microlaryngobronchoscopy; one child had a type 1 laryngeal cleft that was repaired.

Children were followed up for a mean 5 years 0 months (range 1y–7y 7mo). Complete resolution of dysphagia with normal drinking of thin fluids was seen in six children; one child had essentially resolved but would cough if they drank quickly; three children had improved in that they tolerated reduced thickness; five children showed no improvement at all (including two who required a gastrostomy for liquids). In eight children there were no longer any respiratory issues, but seven still had intermittent chest infections or symptoms. The outcome could not be predicted by age at diagnosis, severity of videofluoroscopic finding, or initial level of thickener required.

Table 1: Patient characteristics of 15 children with confirmed isolated bulbar palsy

Patient	Age at firstfeeding symptoms	Age at diagnostic VFSS	Sex	Presenting respiratory symptoms	Aspiration symptoms reported	Clinical assessment of swallow
1	0–6mo	6mo	M	Wheeze	Coughing drinking	No aspiration noted
2	0–6mo	9mo	M	Persistent wet cough	Coughing drinking	Silent aspiration suspected
3	0–6mo	10mo	M	Persistent dry cough	Coughing drinking	Silent aspiration suspected
4	7–12mo	1y 5mo	M	Wheeze	Coughing drinking	Silent aspiration suspected
5	0–6mo	1y 10mo	M	Persistent wet cough	Coughing drinking	No aspiration noted
6	0–6mo	2y 0mo	M	Wheeze	Coughing drinking	Silent aspiration suspected
7	7–12mo	2y 4mo	F	Persistent dry cough Wheeze	Coughing drinking	No aspiration noted
8	0–6mo	2y 5mo	M	Recurrent LRTI	Coughing drinking	Refused to drink
9	0–6mo	2y 5mo	M	Persistent wet cough Wheeze	Coughing drinking	Overt aspiration
10	0–6mo	2y 6mo	F	Persistent wet cough	Coughing drinking	Silent aspiration suspected
11	None	2y 8mo	F	Persistent wet cough	None	Silent aspiration suspected
12	>12mo	2y 9mo	M	Persistent wet cough	Coughing drinking	Silent aspiration suspected
13	0–6mo	4y 2mo	F	Recurrent LRTI	Coughing drinking	No aspiration noted
14	0–6mo	4y 7mo	M	Persistent wet cough Recurrent LRTI	Wet voice	No aspiration noted
15	>12mo	4y 11mo	M	Wheeze	Coughing drinking	Silent aspiration suspected

VFSS, videofluoroscopic swallow study; LRTI, lower respiratory tract infection.

Table 2: Results of the 15 children with confirmed isolated bulbar palsy

Patient	Age at diagnostic VFSS	Main first VFSS finding	Recommended management	Age at last VFSS	Main last VFSS finding	Recommended management	Respiratory progress	Resolved
1	6mo	Penetration level 3	Cooled boiled water	5y 4mo	Penetration level 2, no aspiration	Level 2	8y 1mo. Still has LRTI. Level 2	No
2	9mo	Silent aspiration level 2, penetration purees	Level 3	–	–	–	1y 9mo. No LRTI. Symptom free. Thin fluids	Yes
3	10mo	Silent aspiration level 2	Level 3	2y 10mo	Penetration level 2, no aspiration	Level 2	6y 3mo. No LRTI. Symptom free. Thin fluids since age 3y 9mo	Yes
4	1y 5mo	Silent aspiration level 3	Nasogastric feeding, tastes of breastmilk and purees	2y 8mo	Aspiration level 3	Gastrostomy for fluids, level 4	3y 2mo. No LRTI. Level 4 plus gastrostomy fluids	No
5	1y 10mo	Silent aspiration level 3	Level 3	5y 7mo	Safe on level 2 (not tested on thin)	Level 2	8y 2mo. Still LRTIs. AZM prophylaxis. Sterile water	No, but improvement
6	2y 0mo	Silent aspiration level 2	Level 3	5y 10mo	Silent aspiration level 2	Level 3	9y 4mo. Recent LRTI. On level 2	No, but improvement
7	2y 4mo	Overt aspiration thin fluid	Level 3	7y 8mo	Silent aspiration thin fluid	Sterile water	9y 8mo. No LRTI. Symptom free. Thin fluids	Yes
8	2y 5mo	Penetration level 3, no aspiration	Level 3	–	–	–	7y 3mo. No LRTI occasional wet cough. AZM prophylaxis. Gastrostomy feeds for poor growth. Normal PFTs. Thin fluids	No, but improvement
9	2y 5mo	Silent aspiration level 2	Level 3	3y 11mo	Aspiration solids	Gastrostomy for fluids, tastes of solids	6y 8mo. Occasional mild LRTI. AZM prophylaxis. Gastrostomy fed, some solids orally	No
10	2y 6mo	Silent aspiration thin fluid	Level 2	3y 7mo	Safe on thin	Thin fluids	7y 8mo. No LRTI. Symptom free. Normal PFTs. Can cough if drinks quickly	Yes – partial
11	2y 8mo	Silent aspiration level 2	Level 3	5y 1mo	Safe on thin	Thin fluids	8y 6mo. No LRTI. Symptom free. Using straw – went to normal thin fluids. Type 1 cleft injected 3y 5mo	Yes
12	2y 9mo	Silent aspiration level 2	Level 3	5y 11mo	Penetration level 2, no aspiration	Level 3	8y 1mo. No LRTI. Symptom free. AZM prophylaxis. Level 3	No
13	4y 2mo	Penetration level 2, no aspiration	Level 3	–	–	–	8y 0mo. No LRTI. Wet cough. Normal PFTs. Level 3	No
14	4y 7mo	Silent aspiration level 2	Level 3	–	–	–	10y 8mo. Had been well for 2 years but single LRTI. Thin fluids	Yes
15	4y 11mo	Silent aspiration level 2	Level 3	5y 8mo	Penetration level 2, no aspiration	Level 2	8y 2mo. No LRTI. Symptom free. Normal PFTs. Thin fluids since 7y 10mo	Yes

First diagnostic VFSS, last VFSS if had more than one done, swallow management, and follow-up data. Old nomenclature for International Dysphagia Diet Standardisation Initiative level 2 was syrup, level 3 custard, and level 4 pudding thickness.[5] VFSS, videofluoroscopic swallow study; LRTI, lower respiratory tract infection; AZM, azithromycin; PFT, pulmonary function test.

DISCUSSION

We presented 15 children with dysphagia due to a bulbar palsy in the absence of other neurological causes or impaired neurodevelopment; one also had a type 1 laryngeal cleft. Although nine children with congenital bulbar palsy causing dysphagia were included in a study of 59 patients who had gEMG,[3] our paper is the first to detail management and outcomes of children with isolated bulbar palsy. There are a number of case series of neurologically typical children with chronic aspiration causing respiratory symptoms, and it is quite possible a proportion of them would have had an isolated bulbar palsy as they had not been tested.[7–10]

Knowing the underlying cause can be helpful for some parents coming to terms with the difficult fact of not being able to allow their child to drink normally, something which can cause significant parental concerns and stress.[11] The diagnosis does not alter management, which principally comprises use of thickened fluids (in 12/15 children) which can be quite challenging, especially as all bar one required level 3 thickener. Two children required a gastrostomy, one after initial nasogastric feeding and one after level 3 thickener. One child had cooled boiled water as they were aspirating level 3 thickener and that was preferred over tube feeding. Some are also given alternative drinking strategies such as using a straw, a slow flow teat, chin tuck position, sitting upright, etc.[1]

Prognosis is of primary concern to parents/carers. It is encouraging that 6 out of 15 resolved completely and had a normal swallow at ages ranging from 1 year 9 months to 10 years 8 months (mean 7y 6mo), and 8 out of 15 no longer had respiratory infections or any symptoms. For comparison, 6 out of 8 with a normal gEMG resolved completely with a normal swallow at ages 2 years 8 months to 12 years 5 months (mean 9y 9mo). Parents should be informed that clinical progress is likely to be slow, children follow their own trajectory, and there is no clinical marker to indicate outcomes. There are no series of children with an isolated bulbar palsy to compare our data with. However, one series of 13 typically developing infants presenting with respiratory symptoms found all had resolved in just 3 to 9 months after starting thickened feeds, which is surprisingly fast from our experience.[7] A larger series of 50 otherwise typically developing infants found the probability of resolution was 46% at 6 months, 64% at 1 year, and 76% at 2 years.[10] Another series of 19 children with isolated dysphagia and without apparent risk factors found on follow-up that 3 out of 14 still had swallowing problems aged 9 to 14 years, whilst 11 out of 14 had resolved at 8 months to 10 years of age (mean 3y 2mo).[9]

It would be interesting to repeat the gEMG to study change over time, particularly in those whose dysphagia had resolved to see if the gEMG had also improved. However, we are not certain many parents would agree to put their child through the procedure when they are now symptom free and drinking normal fluids. There is no clinical justification, and it is uncertain how an ethics committee would regard an invasive procedure for research purposes alone in young children. Equally, repeating the gEMG in those who still had dysphagia is not justified as it will not change management.

There was also no indicator on VFSS as to which children might have a bulbar palsy, although the eight children with a normal gEMG were older at presentation, with 4 out of 8 aged 5 years and above (0/15 over 4y in isolated bulbar palsy group). Despite these patients having a bulbar palsy, their swallowing did not present with characteristics seen in children with more generalized neurological impairments;[12] specifically our cases did not have oral phase difficulties and residue, and less than half had a delayed swallow.

We cannot estimate the prevalence of an isolated bulbar palsy as not all children with dysphagia are sent for a gEMG. Furthermore, there is a false negative rate for gEMGs which we cannot estimate since there is no other way of diagnosing isolated bulbar palsy. The policy of our respiratory team now is to refer children to a neurologist for neurological evaluation and consideration of further investigations who have obvious neurological/neurodevelopmental issues, if they deteriorate over time, require level 3 thickener for a safe swallow, or remain symptomatic despite use of the thickener (assuming good adherence). Some parents prefer their child does not undergo a gEMG as although it is usually well tolerated,[13] some children find it challenging. Additionally, the diagnosis of an isolated bulbar palsy will not alter management nor give a definite prognosis. Not all children underwent a microlaryngobronchoscopy, but the fact one child had both a type 1 laryngeal cleft as well as a bulbar palsy has made us consider the recommendation that all should have a microlaryngobronchoscopy,[2] even though it requires a general anaesthetic. Certainly, a multidisciplinary approach to children with dysphagia is required.

In conclusion, we presented a series of children with an isolated bulbar palsy as a cause of their oropharyngeal dysphagia who presented with respiratory symptoms, and also a group in whom isolated bulbar palsy could not be diagnosed because of a normal gEMG. Isolated bulbar palsy in children is a distinct entity with a cause that remains elusive. Whilst knowing the underlying diagnosis has limited practical use, many parents wish to understand why their child has a significant swallowing problem so investigation with a gEMG should be considered.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

SUPPORTING INFORMATION

The following additional material may be found online:

Table S1: Patient characteristics of the eight children with a normal EMG

Table S2: Results on the eight children with a normal EMG

Table S3: Characteristics of swallow on initial diagnostic video fluoroscopic swallow study in 15 children with isolated bulbar palsy

Appendix S1: Methods

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