Bulbar and Pseudobulbar Palsy

Synonyms: ‘bulbar palsy’ - lower motor neurone dysarthria, neuromuscular dysarthria, atrophic bulbar paralysis; 'pseudobulbar palsy' - upper motor neurone dysarthria, spastic dysarthria

The components of normal speech

Speaking is a voluntary task which is taken for granted but is a highly specialised activity. In order to speak, the following parts of the oral cavity need to be used:

- Larynx
- Pharynx
- Palate
- Tongue
- Lips

Along with this, controlled expiration is required, so that air can be released at the appropriate speed and in appropriate amounts.

Neurological control of normal speech

The above structures required for speech are controlled by the nervous system. Corticobulbar tracts from both of the motor cortices send signals down to the nuclei of the following nerves:

- Vagal nerve
- Facial nerve
- Hypoglossal neve
- Phrenic nerve

The motor aspects of speech, like other movements, are also influenced by the extrapyramidal system via the basal ganglia and the cerebellum. There is ongoing research into which areas of the cortex are involved in speech and especially in recovery of speech after a stroke. [1]

Phonation and articulation

Speech has two elements: phonation and articulation.

- **Phonation**: the production of sounds, a result of the vocal cords in the larynx.
- **Articulation**: contractions of the muscles of the various other structures involved in speech, i.e. the pharynx, palate, tongue and lips. These muscle contractions change the vocal sounds from the larynx, thus resulting in noises recognised as words.

Larynx - produces vowels and some consonants.

Disorders of articulation [2]

This is also called dysarthria or anarthria. There are several causes:

- Bulbar palsy
- Pseudobulbar palsy
- Cerebellar-ataxic
- Hypokinetic
- Hyperkinetic
Bulbar palsy

Description
Bulbar relates to the medulla. Bulbar palsy is the result of diseases affecting the lower cranial nerves (VII-XII). A speech deficit occurs due to paralysis or weakness of the muscles of articulation which are supplied by these cranial nerves. The causes of this are broadly divided into:

- Muscle disorders.
- Diseases of the motor nuclei in the medulla and lower pons.
- Diseases of the intramedullary nerves of the spinal cord.
- Diseases of the peripheral nerves supplying the muscles.

Importantly, these lesions do not affect speech in isolation. The bulbar nerves also innervate muscles involved in swallowing and facial muscles.

Bulbar palsy is sometimes also classified as non-progressive or progressive. Non-progressive bulbar palsy is an uncommon condition of uncertain aetiology and there are few reports of it in the literature.[3] Progressive bulbar palsy can occur in children or adults and form a spectrum of severity, based around the common feature of bulbar dysfunction and motor neurone degeneration. Genetic abnormalities have been identified in some cases presenting in childhood. Brown-Vialetto-Van Laere and Fazio-Londe syndromes are the most recent childhood forms of progressive bulbar palsy to be genetically defined.[4]

Presentation

- Lips - tremulous.
- Tongue - weak and wasted and sits in the mouth with fasciculations.
- Drooling - as saliva collects in the mouth and the patient is unable to swallow (dysphagia).
- Absent palatal movements.
- Dysphonia - a rasping tone due to vocal cord paralysis; a nasal tone if bilateral palatal paralysis.
- Articulation - difficulty pronouncing r; unable to pronounce consonants as dysarthria progresses.

If the pathology progresses then speech becomes slurred and eventually becomes indistinct. There may also be neurological deficits in the limbs - eg, flaccid tone, weakness with fasciculations.

Aetiology[5]
There is a wide range of causes. The following list is not exclusive:

- Diphtheria.
- Poliomyelitis.
- Motor neurone disease - eg, progressive bulbar palsy (features of pseudobulbar palsy may also be present).
- Syringobulbia.
- Cerebrovascular events of the brainstem.
- Brainstem tumours.
- After radiotherapy for nasopharyngeal carcinoma.
- After surgery for acoustic neuroma.
- Guillain-Barré syndrome.[6]
Pseudobulbar palsy

Description
Pseudobulbar palsy results from disease of the corticobulbar tracts. Bilateral tract damage must occur for clinically evident disease as the muscles are bilaterally innervated.

Presentation
- Tongue - paralysed; no wasting initially and no fasciculations; 'Donald Duck' speech; unable to protrude.
- Palatal movements absent.
- Dribbling persistently.
- Facial muscles - may also be paralysed.
- Reflexes - exaggerated (eg, jaw jerk).
- Nasal regurgitation may be present.
- Dysphonic.
- Dysphagic.
- Emotional lability may also be present.

There may also be neurological deficits in the limb - eg, increased tone, enhanced reflexes and weakness.

Aetiology
- Cerebrovascular events - eg, bilateral internal capsule infarcts.
- Demyelinating disorders - eg, multiple sclerosis.
- Motor neurone disease.
- High brainstem tumours.
- Head injury.
- Neurosyphilis.

In motor neurone disease it is common to see both bulbar and pseudobulbar palsies.

Investigations
New developments in technology have led to the use of neurophysiological investigations to assess various aspects of speech dysfunction. These include electromagnetic articulography (EMA), electropalatography (EPG) and pressure-sensing EPG. Other tests will depend on the suspected underlying cause but will involve routine blood tests, imaging of the brain and brainstem (either CT scan or MRI) and electromyography.

Management
- All patients should be referred to neurologists. Patients will need admission if dysphagia is present or symptoms are rapidly progressive.
- Treatment will be directed to the underlying cause.
- Postural changes can help with drooling of saliva and may prevent aspiration.
- Supportive measures may include baclofen for spasticity, anticholinergics for drooling, treatment of aspiration pneumonia if it occurs and attention to nutrition - eg, enteral feeding.
- Management should involve speech and language therapists and the dietician.
- Genetic analysis may be appropriate for cases presenting in childhood.

Complications
- Poor dentition.
- Poor nutrition.
- Psychological dysfunction.
- Progression of underlying disease.
Prognosis

This depends on the underlying cause.

Further reading & references

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