Sialorrhea in cerebral palsy

Introduction

Sialorrhea, also called hypersalivation or ptyalism refers to involuntary drooling of saliva as a result of limitations in a person's ability to control and swallow oral secretions. A proper swallowing reflex is essential for swallowing of saliva. This complex process is mediated by orofacial muscular systems and involves a series of sequential reflexes and coordinated movements of the muscles of mandible, lips, tongue, pharynx, larynx and oesophagus. Drooling is considered to be normal in typically developing children up to 24 months. It may be tolerated up to 4years, but persistence after 4 years is considered pathologic (1). It often persists in children with neurologic disorders including neuromuscular incoordination of swallowing and intellectual disabilities. Generally, sialorrhea may be arise from excess production (primary) or due to poor control (secondary). From a clinical point of view, sialorrhea may be further be classified into anterior and posterior. *Anterior drooling* is defined as saliva spilled from the mouth that is clearly visible. *Posterior drooling* occurs when saliva spills through the oropharynx and into the hypopharynx.

The prevalence of sialorrhea in cerebral palsy (CP) from various studies has varied between 10% and 58% in children/youth with CP (2). Aetiology in CP is often multifactorial and may be the result of limited oromotor control as a result of muscle incoordination and intraoral sensory perception difficulties rather than excessive salivation, and dysphagia. Factors correlated with sialorrhea in CP include; difficulties in formation of food bolus, inefficient labial sealing, suction disorder, increased food residue, difficulty controlling the lips, tongue and mandible, reduced intraoral sensitivity, reduced frequency of spontaneous swallowing, oesophageal dysphagia, and dental malocclusion. Other factors all common in CP influence the presence and severity of sialorrhea: Open mouth position, inadequate body posture, particularly of the head, intellectual disability, emotional state, and degree of concentration (2). In a study by Reid *et al*, they analysed the predisposing factors of sialorrhea in children with CP, aged 7 – 14 years which included: non-spastic types, spastic quadriplegic CP, absence of cervical control, severe difficulty in gross motor coordination/function, epilepsy, intellectual disability, lack of speech, open anterior bite and dysphagia (3).

Pathologic drooling can have significant medical and psychosocial impact on the child, which increases disability and reduces quality of life for the child and work overload for the caregivers. The medical concerns include; chronic aspiration arising from posterior pooling that can result in recurrent infections and progressive lung disease. The presence of saliva on the chin leads to frequent wiping, causing skin irritation and breakdown, and predisposes to oral and perioral infections, tooth decay, difficulty with hygiene, dehydration and electrolyte imbalances and unpleasant odour. Psychosocial concerns of anterior drooling include need for frequent clothing changes, or for child to wear a bib. Spilled saliva may damage books, computers, toys and other equipment, and cause a spray from the mouth while talking. This may cause social embarrassment to the child/youth, their caretakers and siblings, and may lead to isolation and low self-esteem. This may affect their socio-emotional development.

Loss of saliva may impair removal of gastric acid reflux into the oesophagus, which can perpetuate oesophageal dysmotility and oesophagitis and hence risk of gastroesophageal reflux (GERD). On the other hand, GERD may result in increase in salivation to protect the upper

oesophagus. However, treatment of GERD has not been associated with reduced sialorrhea in CP (4).

Sialorrhea assessment

It is necessary to quantify the frequency and severity of sialorrhea. There is currently no consensus regarding assessment. Discussion with providers from multiple disciplines is recommended. Medical assessment should emphasise on history of medications use and aspiration. Systematic clinical examination should be done and includes position of head during swallowing and trunk posture relative to head position. Orofacial examination should cover dentition, maxillary deformity, lip closure, oral hygiene, upper airway pathology, perioral skin and mucosa, orofacial-lingual dyspraxia. Motor/Oromotor assessment should cover head control, positioning, mouth closure, occlusion, lip seal, oral sensorimotor evaluation, swallow on demand, ability to wipe own saliva. Also asses hydration, lower respiratory system, a neurologic assessment covering; craniofacial control, posture, impact of medicines, epilepsy, developmental age equivalent, and asses for GERD, and presence of allergy. Social evaluation should cover behaviour and intrinsic motivation and child's self-management skills, impact of sialorrhea and importance of saliva control to family.

Drooling can be assessed quantitatively with a variety of tools for severity and frequency, as well as impact on the child and family. The development of validated tools for assessment remains a challenge. Both subjective and objective measures have been used to asses impact and severity of drooling. Subjective tools include: 'subjective evaluation by family/carer, Teacher Drooling Scale, Drooling Severity and Frequency Scale, Visual Analogue Scale (VAS), Drooling Impact Scale (DIS), number of bibs, and frequency of clothing changes. These are filled up by patients or their caregivers and express their qualitative and quantitative impressions of the severity and impact of sialorrhea. Objective methods include measurement of salivary flow and direct observation of salivary loss. Examples of objective tools used include: Drooling quotient, Sochaniwskyj's technique, Thomas- Stonnel, Greenberg scale, number of swallows in a given time, weight of saliva collected on cotton in 5 minutes etc. Quantification can aid in gauging response to interventions.

Differentiation between anterior and posterior drooling is important. They may appear independently or may co-exist. Most often, clinical information such as repeated episodes of pneumonia, repeated antibiotic courses for respiratory reasons, evidence of chronic inflammatory lung disease, and significant need for suctioning are used as indicators of posterior drooling. Additional investigations to consider include salivagram and fiberoptic endoscopic evaluation of swallowing; however, they may not always be necessary or appropriate.

Management:

Care pathways have been described by consensus. A number of treatment strategies are available although there is no clear consensus as to which are safe and effective. Hierarchal approach is taken, from the least invasive therapies to the more invasive ones. Goals of treatment target: 1) improvement of oromotor control of secretions; 2) enhancement of a child's ability to behaviorally manage secretions; and 3) reduction of saliva production or rerouting of salivary flow. 4) Improving quality of life for the patient and caregivers, and reduction of burden experienced by caregivers. When possible, a multidisciplinary team approach is recommended, progressing from conservative to more invasive treatments until saliva control

is improved and side effects, if present, are manageable. Complete control is often not possible. Rehabilitation may consist of oro-motor therapy, biofeedback, or behavioural interventions; however, few studies have assessed their effectiveness. Medications use has been limited by side effects. Botulinum toxin intraglandular injection has been shown to be effective. Surgical interventions may be used but may not be curative. There is a lack of consensus regarding which interventions are most effective for children with CP. All of the strategies that follow may be appropriate for anterior drooling. However, oromotor and orosensory strategies, behavioural strategies, and oromotor appliances are not recommended for posterior drooling. Duct relocation is contraindicated for posterior drooling.

Specific management approaches:

- **Optimize conditions -** Optimize positioning and medical management of factors that affect drooling. Consider whether medications being used for other conditions, such as epilepsy, baclofen, etc are increasing drooling.
- **Oromotor and orosensory strategies -** Active and passive exercises as well as sensory applications are widely used by clinicians, although there is no agreement about the theoretical basis and effectiveness of these interventions. In a systematic review by Dias in 2016, training of sensory awareness and oromotor skills by a speech therapist was found to be the most effective strategy in CP children (2). However, these approaches can be time consuming. Success depends on the child's intellectual capacity. No adverse effects are reported. It has long lasting effect. In a randomised controlled trial by Inal *et al* in 2017, to assess the role of functional chewing training on tongue thrust and drooling in children with CP, a significant difference was found between the intervention group versus the control group, suggesting benefit of this intervention (5).
- **Behavioural strategies -** Multiple types of behavioural procedures have been shown to be effective (low level evidence). Body modification through biofeedback has been used. Based on the monitoring of the target muscle group for electromyographic (EMG) stimulation. When the muscle contracts, EMG informs of the change in muscle activity through acoustic or light signals. Thus, the patient can consciously correct or improve certain components of swallowing. Selection and success depend on the ability of the child to comply and often requires on-going effort for maintenance of effect. No adverse effects are reported.
- **Oral appliances -** Compliance can be challenging and nose breathing must be possible for the child wearing the appliance. Children with seizure disorders may be at risk for oral injury. There is some low-level evidence that oral appliances may be effective.
- **Anti-cholinergic agents which inhibit salivary secretion –** Inhibit the parasympathetic input in the glands, hence inhibiting salivary flow. Drugs may be administered as oral, cutaneous or transdermal or sublingual. There is limited evidence of their effectiveness. Glycopyrrolate, scopolamine (also known as hyoscine), benzhexol and benztropin are the most commonly used agents internationally. Use has been limited by side effects such as excessive thickening of secretions, urinary retention, constipation, headache, blurred vision and behavioural disturbance. In a prospective study done in 2019 comparing role of Benzhexol, glycopyrrolate, and scopolamine in reducing drooling, they were all shown to reduce drooling, but improvement was offset by adverse side effects and glycopyrrolate performed best with fewer side effects compared with benzhexol and scopolamine (6). Atropine Sulphate antagonises muscarinic receptors in salivary glands and leads to reduced saliva production. Its effect was evaluated in a non- controlled clinical trial by Dias *et al.* It was shown to have good clinical response combined with good safety profile (7). It is

also inexpensive. Trihexyphenidyl has been used in dystonic CP or selected cases (2). Anticholinergics have the risk of possible exacerbation of GERD and oesophagitis.

- **Intraglandular Botulinum toxin injections to the submandibular +/- parotid glands** – Botulinum inhibits release of acetylcholine from cholinergic nerve terminals, thereby reducing salivary secretion and sialorrhea. Injections are often considered after inadequate response to anti-cholinergic treatment. They can be effective in reduction in reduction in secretion of saliva and in drooling. However, it needs to be repeated regularly, often at 6-month intervals, and responsiveness may diminish over time. Botulinum toxin is most often injected using ultrasound guidance for assistance. Procedure requires some sedation or anaesthesia. Varying doses have been reported with botulinum toxin A being the most frequently used type. One study suggested initial total doze of 80U, and could be increased to 120U if response is insufficient (8). Several prospective cohort studies have supported efficacy of botulinum A toxin on reducing the severity of drooling and frequency of respiratory infection in CP children with posterior drooling (9, 10). It has been shown to be safe, but potential side effects include irritation at the injection site, pain, hematoma, dry mouth, thickened secretions or problems with chewing and swallowing from diffusion to the surrounding submental muscles thus, increasing aspiration risk. More side effects should be anticipated in patients with 2 gland injections, than submandibular injections alone or after repeated gland injections in the same individual (11). Caution should be taken for patients with dysphagia, which may worsen (11, 12).
- **Surgical intervention**. Surgery is usually reserved for patients with profuse, persistent anterior drooling, continued symptoms despite maximal conservative or pharmacological treatment, and patients with posterior drooling who have chronic aspiration and/or recurrent respiratory infections. Surgical procedures may include duct ligation or rerouting, sublingual or submandibular gland excision, and varying combinations of these procedures. Submandibular gland resection has been shown to reduce salivary flow by up to 80% in some cases, but success and caregiver satisfaction are variable (13, 14). Side effects are usually minimal, but include xerostomia, wound infection, dysarthria, and dental malocclusion. Duct recanalization can occur. In a study that reviewed CP cases that had undergone unsuccessful submandibular gland duct surgery for drooling, they reported some cases of failure, which was attributed to coexisting non- surgical complications such as dysarthria and dental malocclusion (15).
- **Sensory level electrical stimulation –** Electrical stimulation has been shown to improve sialorrhea in adult patients with neurologic disease. In a study done to evaluate the role of electrical stimulation in paediatric CP patients with dysphagia, they suggested that this may be an effective modality to improve oropharyngeal symptoms, severity of dysphagia and dysphagia level in paediatric patients (16).
- **Repeated muscle vibration –** This technique was assessed in one study. They used proprioceptive impulse to activate fibres 1a reaching the somatosensory and motor cortex. It was applied using an impulse that was applied to the chin symphysis for 30 minutes for 3 consecutive days. Its effect on drooling was assessed and was found to be safe and effective. The vibration was shown to improve swallowing mechanisms and favoured acquisition of the maturity of the oro-motor control in children with CP (17).

Longitudinal Management

Whether or not an intervention is utilized, the psychosocial and medical effects of drooling must be monitored longitudinally. If an intervention is pursued, regular systematic

monitoring of the child and caretaker for indications of efficacy and potential side effects is imperative.

Saliva is responsible for mechanical cleaning and protective functions which is essential to the maintenance of oral health. There is a potential risk of increased risk of dental caries with reduction in salivary flow rate. In a study done to evaluate this in CP children put on anticholinergics, botulinum A toxin, or surgery, higher rates of dental spots were observed in children that had undergone surgical procedures (18).

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