Congenital brachial plexus palsy

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Abstract

Congenital brachial plexus palsy (CBPP) occurs secondary to traumatic injury to the brachial plexus in the perinatal period. It is a commonly encountered problem on the postnatal wards with the potential for significant morbidity. Previously termed 'obstetrical brachial plexus palsy', recent evidence suggests that it may occur independently of the actions of the birth attendant in some cases. Significant risk factors include shoulder dystocia, macrosomia, instrumental delivery and prolonged second stage of labour. Prognosis is variable and dependent on the type and level of nerve injury with total plexus palsies having the worst prognosis. Approximately two thirds of patients will have complete and spontaneous recovery by 6 months of age. However in those with residual defects, secondary complications may develop. It is important to diagnose it early and manage appropriately due to the risk of long term neuromuscular dysfunction and deformities in the affected arm in some cases.

Management following recognition involves, prompt paediatric review and exclusion of associated injuries such as bony fractures. Urgent and regular physiotherapy input is crucial in preventing complications and monitoring progress. Primary surgical repair may be indicated in cases that do not or are unlikely to recover and prompt and early referral to a specialist centre is recommended in these difficult cases. Secondary surgery may also be indicated in those who develop complications. This review details the pathophysiology, risk factors and classification of CBPP. It suggests a systematic approach to the diagnosis of CBPP and gives a framework for understanding its management and prognosis.

Keywords birth trauma; brachial plexus; brachial plexus palsy; congenital; obstetric

Introduction

Congenital brachial plexus palsy (CBPP) results in a flaccid paresis of the upper limb at birth with a greater passive range of limb movement than active. It is usually unilateral but can be bilateral. It was first recognised as having an obstetrical origin by Smellie in 1768 following his clinical description of an infant with transient bilateral arm paralysis following a prolonged and difficult delivery and hence has also been referred to as obstetrical brachial plexus palsy, reflecting it's presumed pathophysiology. CBPP is generally believed to be secondary to traumatic injury to the brachial plexus in the perinatal period and is readily apparent at or shortly after birth. The current reported incidence of CBPP in Europe and the United States is 0.42–3.2/1000 live births. The clinical presentation varies according to the extent of the damage from mild limitation of movement of the affected limb to complete flaccid paralysis of entire upper limb – 'flail arm' with or without Horner's syndrome (ptosis, miosis). Although most cases have full functional recovery, it can result in persistent disability which can be a source of medical litigation claims.

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Pathophysiology

CBPP occurs secondary to trauma to the brachial plexus, resulting in damage to some or all of its nerve roots (C5-8, T1). The four types of nerve injury described are avulsion, rupture, neuroma and neuropraxia. Avulsion and rupture are the most serious forms of nerve injury whereas post traumatic neuroma formation and neuropraxia have a better prognosis. Nerve avulsion occurs when the nerve is torn away at its point of attachment to the spinal cord and has no potential for recovery. Nerve rupture indicates stretching with subsequent tearing of the nerve distal from this site of attachment. This may be partial or complete. A traumatic neuroma is a benign proliferation of Schwann cells and axons that develop at the proximal end of a severed or injured nerve. This can be considered as 'scar tissue' and may lead to nerve compression. Neuropraxia occurs secondary to nerve stretching and can be described as a transient loss of motor and sensory function due to blockage of nerve conduction. In these cases full recovery of nerve function is often achieved within 2–3 months.

The nerve trauma in CBPP has traditionally been reported as occurring due to excessive downward traction on the brachial plexus at the point of delivering the anterior shoulder by the birth attendant, particularly in cases of difficulty in delivering the shoulders. It has also been recognised as occurring secondary to hyperextension of the arms following breech vaginal delivery. However, recent studies have suggested that other mechanisms may be involved independent of the actions of the midwife or obstetrician, as up to 1/3rd of cases of CBPP occur in the absence of recorded shoulder dystocia and/or involve the posterior shoulder. Other proposed mechanisms include a failure of normal rotation of the fetal shoulders into an oblique position or persistent impaction of the posterior shoulder against the sacral prominence prior to delivery.

Recognised risk factors for CBPP include shoulder dystocia and macrosomia (birth weight more than 4.5 kg). Instrumental delivery, maternal diabetes, prolonged second stage of labour, increased maternal BMI and a previous infant with CBPP are also known to increase risk. Intrauterine torticollis is also a risk factor. However, it is worth noting that some neonates with CBPP will not have any recognisable risk factors and the implementation of training for midwifery/obstetric staff in preventing shoulder dystocia has had a variable impact on CBPP incidence.

Diagnosis

The clinical features of CBPP are recognisable from or shortly after birth and vary according to the level and degree of nerve root damage. The most common type of brachial plexus palsy

involves the upper part of the plexus only (C5-7) and is frequently referred to as Erb's palsy. It presents clinically as internal rotation and adduction of the shoulder, elbow extension with flexion of the wrist and extension of the fingers. This is classically described as a 'waiter's tip' posture and has greater potential for recovery than other forms of CBPP.

Less commonly, the entire brachial plexus is involved (C5-8, T1). This form of injury has a poor prognosis and is recognised by complete flaccid paralysis of the arm – 'flail arm' with a 'clawed hand' appearance. This can be associated with Horner's syndrome following concurrent damage to the sympathetic chain.

Damage to the lower brachial plexus alone (C8, T1) is very rare, accounting for only 2% of all reported CBPP and is also referred to as Klumpke's Palsy. In these cases, proximal muscle function is spared with poor hand grip being the primary clinical feature. The forearm may be supinated with hyperextension of the wrist and fingers (claw hand) with preservation of shoulder and elbow function.

Classification and natural history

The most common clinical classification system used in CBPP is the Narakas score which identifies the level of nerve root involvement according to clinical features. The extent of the nerve roots affected influences the long-term outcome, with upper plexus palsies generally having a better prognosis (Table 1).

The severity of the nerve injury also influences the prognosis with nerve rupture and avulsion resulting in poorer outcomes. Other classification systems based on the physiological nature of the brachial plexus injury have also been used. Sunderland in 1974 classified CBPP according to increasing severity of nerve injury with type 1 representing neuropraxia (good prognosis) and type 5 representing complete nerve transaction (no spontaneous recovery).

Previous studies had reported excellent spontaneous recovery rates in CBPP, up to 75–95%. However, a recent evidence based review by Foad et al in 2009 found that only two thirds of infants with CBPP achieve complete recovery of function by 6 months of age without intervention and therefore care should be taken not to be over-optimistic when counselling parents. Narakas type I and II palsies are likely to have a better recovery rate compared to type III and IV. 20–33% patients can have residual deficits and are therefore at risk of long term complications including contractures of shoulder and elbow, arm hypoplasia, pseudowinging of the scapula, altered sensation in the arm, muscular weakness, dislocation and long term psychological and social difficulties. Therefore, careful monitoring and timely surgical intervention may be required.

Associations

CBPP may be present in association with other birth related injuries; in particular fractures of the clavicle and humerus. Wall et al in 2014 identified that brachial plexus palsy in the presence of clavicular fracture may have an improved prognosis compared to isolated brachial plexus palsy, as clavicular fracture may ease the delivery of the anterior shoulder. Other birth injuries associated with CBPP include sternocleidomastoid injury resulting in contraction or mass, facial or phrenic nerve palsy, transient hypoglossal and recurrent laryngeal injuries. Therefore, it is important that all the infants with CBPP are fully examined to exclude the above associations and undergo appropriate radiological investigations.

Differential diagnosis

The differential diagnosis for upper limb paralysis in the newborn includes epiphyseal separation of the humeral head and bony fracture of the clavicle or humerus with associated pseudoparalysis. Spinal injury and cervical cord lesions, congenital varicella, arthrogryposis and rare tumors such as myofibromatosis affecting the brachial plexus should also be considered.

Classification of CBPP and associated estimates for incidence and prognosis

Narakas group	Other name	Roots involved	Incidence	Clinical features	Complete recovery
I	Upper Erb's palsy	C5-6	73%	Shoulder adducted and internally rotated, elbow extended, forearm pronated	3 months 59%, 6 months 65%
II	Extended Erb's palsy	C5, 6, 7		As above + wrist flexed/fingers extended in 'waiters tip position'	
III	Total palsy	C5, 6, 7, 8, T1	25%	Complete flaccid paralysis of entire upper limb – flail arm	3 months 0%, 6 months 14%
IV	Total palsy + Horner's	As above		As above + Horner's	
	Klumpke's Palsy	C8-1	<2%	Forearm supinated, wrist and fingers hyperextended – 'claw hand'. Good elbow and shoulder function	<50%

Table 1