

## CLINICAL CASE REPORT

# Paediatric neurological melioidosis: a rehabilitation case report

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## ABSTRACT

**Context:** Melioidosis is a rare condition, endemic to northern Australia and south-east Asia, caused by an infection from the bacteria *Burkholderia pseudomallei*. The largest epidemiological review to date describes 540 cases of melioidosis seen at Darwin Hospital, in northern Australia, over a 20-year period. Of these, 14 (less than 3%) presented with neurological manifestation, with three deaths. Reports of paediatric cases of melioidosis are rarer. In a review of paediatric cases in northern Australia only eight cases were identified in 10 years. Three of these patients presented with neurological melioidosis, of whom two died in hospital.

**Issues:** Whilst the literature refers to prolonged periods of hospitalisation for survivors, the trajectory of functional recovery and process of rehabilitation has not been described. This is a case report describing a 14-year-old boy who presented to a remote medical post with acute neurological symptoms (vomiting, severe headache, ataxia, cranial nerve VI and VII palsy) and was referred to the tertiary paediatric hospital in Perth, Western Australia. Cranial magnetic resonance imaging showed an extensive infiltrative lesion in the posterior fossa and hydrocephalus. Diagnosis of neurological melioidosis required isolation of the pathogen by brain biopsy through sub-occipital craniotomy. Medical treatment included surgical management of hydrocephalus, parenteral antibiotic treatment with meropenem and then a prolonged course of oral co-trimoxazole, enteral feeding and tonal management with levodopa-carbidopa and botulinum toxin A injections. Associated neurological signs and symptoms (bradykinesia, tremor, dysphagia, aphasia, hypertonia, extropia) required intensive rehabilitation to address functional deficits and to promote independence. The purpose of this case report is to document the functional recovery and rehabilitation process of a paediatric case of neurological melioidosis. Knowledge of the recovery pathway is important to add to the understanding of natural history and treatment of this rare disease.

**Lessons learned:** Occasions of service and functional assessments were recorded prospectively. Inpatient therapy (932 hours, with 934 occasions of service) was delivered across physiotherapy, occupational therapy and speech pathology over 9 months of an



inpatient admission. Initial paediatric functional independence measure (WeeFIM) was 18/126, indicating complete dependence in all physical and cognitive domains. Following 9 months of intensive rehabilitation the WeeFIM was 53/126, indicating significant residual disability. This proved to be a challenge for discharge planning back to a remote region of Western Australia. Paediatric neurological melioidosis can lead to significant disability and long-term dependence, despite the provision of lengthy intensive rehabilitation. This case report highlights the challenges and complexity of the rehabilitation services required to optimise outcomes for this patient and achieve a safe discharge to a remote community where limited support services are available.

**Key words:** Aboriginal health, allied health, Australia, neurological melioidosis, paediatric, rehabilitation.

## Context

### Introduction

Melioidosis is a condition characterised by infection with the bacteria *Burkholderia pseudomallei*, common in groundwater sources during the wet season in tropical locations<sup>1,2</sup>. Infection usually occurs via inoculation, ingestion or inhalation<sup>2,3</sup>. Melioidosis is a rare condition and is endemic in northern Australia and south-east Asian regions<sup>1,2</sup>, with the largest epidemiological review to date emerging from Darwin Hospital in the Northern Territory in Australia. Five hundred and forty cases of melioidosis were prospectively reviewed with variability in the clinical presentation, including pneumonia (51%), genitourinary conditions (14%), skin infection (13%), septic arthritis and/or osteomyelitis (4%). Only 14 patients (3%) presented with neurological melioidosis<sup>1</sup>. The diagnosis of neurological melioidosis is complex and difficult given the variability in clinical presentation<sup>2</sup>. Neurological melioidosis appears to be the rarest presentation and has a high mortality rate<sup>1</sup>. Paediatric melioidosis appears even rarer, with those surviving presenting with long hospital admissions and long-term neurological deficits as a result of their infection<sup>2,4</sup>.

Whilst the literature refers to prolonged periods of hospitalisation and ongoing disability for survivors of neurological melioidosis<sup>4-6</sup>, the trajectory of functional recovery and the rehabilitation required has not yet been described. Knowledge of the recovery pathway is important

to add to the understanding of natural history and treatment of this rare disease. This case report discusses the extended period of rehabilitation required to optimise outcomes in one patient and to highlight the significant degree of residual disability that remained after 9 months and the challenges this presented for discharge to a remote community.

The aim of this paper is to document the functional recovery, rehabilitation pathway and challenges in a single case report of a survivor of paediatric neurological melioidosis from a remote Aboriginal community in Western Australia. This case report is structured using the *CARE guidelines: consensus-based clinical case reporting guideline development*<sup>7</sup>.

Therapist's intervention hours were taken from the hospital statistics database over the nine month period since admission and included physiotherapy, occupational therapy and speech pathology data.

### Patient information

A 14-year-old Aboriginal boy presented for treatment at Jigalong medical post in northern Western Australia with a 2-week history of ataxia and severe headache and 3-day history of vomiting. He had a history of solvent abuse. He was transferred to the closest major paediatric tertiary hospital, more than 1300 km away. At presentation he had lower cranial nerve signs, left-sided facial and limb weakness, dysphasia and was only able to ambulate with the support of two people. He appeared drowsy, although his Glasgow Coma Scale (GCS) was 15, and he remained orientated to



time, person and place. Magnetic resonance imaging (MRI) showed a large infiltrating enhancing lesion with associated mass effect within the brain stem and extending in to the brachium pontis, left cerebellar hemisphere and cerebral peduncles, resulting in obstructive hydrocephalus. He underwent insertion of an extra-ventricular drain to manage his hydrocephalus. Despite this, his clinical presentation continued to deteriorate over subsequent days; his GCS fluctuated between 7 and 12, with the emergence of dysarthria, dysphagia, a dense left hemiplegia and increased muscle tone in his left upper limb, resulting in admission to the intensive care unit. Neurological melioidosis was diagnosed following sub-occipital biopsy of the cerebellar mass lesion identified on cranial MRI. The bacteria were isolated 9 days into his admission and he was treated with IV meropenem followed by a prolonged course of co-trimoxazole (TMP-SMV), in accordance with treatment protocols identified in medical literature<sup>2,8</sup>.

## *Ethics approval*

Informed consent was obtained for publication of this case report as well as ethics approval through the Child and Adolescent Health Service of Western Australia, approval number 2015160QP.

## Issues

### *Clinical findings*

Functional examination at the time of referral revealed that the patient was unable to move independently in bed, unable to sit unsupported and required full assistance from two staff for hoist transfers and all personal care tasks. He was unable to communicate consistently, even with basic yes/no responses. Eye gaze responses were the most effective form of communication; however they were unreliable and limited by fatigue and oculomotor dysfunction. Shared attention and appropriate eye contact with therapists was an encouraging indicator of cognitive preservation. He was unable to swallow safely and required nasogastric tube-feeding.

Neurological examination revealed a dense left-sided hemiplegia with significant spasticity, marked ataxia, bradykinesia, right-sided intention tremor, left exotropia (unilateral cranial nerve VI palsy), left facial weakness (unilateral cranial nerve VII palsy) with protective gag reflex intact. Initial paediatric functional independence measure (WeeFIM)<sup>9</sup> was completed, with a score of 18/126, indicating the need for total assistance in all domains of function (Table 1).

Standardised cognitive assessment was not possible. A valid assessment tool could not be found for this child, whose culture and limited opportunities for formal education reflected his life spent in a remote, isolated and primarily Aboriginal community in Western Australia.

### *Diagnostic assessment*

MRI was repeated throughout admission to monitor disease progression. Common signs often present on MRI included leptomeningeal enhancement, ring-enhancing lesions, oedema, abscess and involvement of the brainstem<sup>6</sup>. This patient developed all of these clinical features throughout his admission, the most prominent of which were the ring-enhancing lesions. Figure 1 illustrates changes as shown by MRI at 3, 6 and 9 months.

### *Therapeutic interventions*

The patient was referred to the acquired brain injury paediatric rehabilitation team for multidisciplinary inpatient rehabilitation in the early stages of his admission. The acquired brain injury team included senior clinicians from paediatrics, paediatric rehabilitation medicine, physiotherapy, occupational therapy, speech pathology, dietetics, nursing, case management, teaching, social work and psychology, with access to Aboriginal liaison staff at Princess Margaret Hospital, which provides tertiary paediatric care to Perth and all areas of the state of Western Australia. A review of the rehabilitation therapists' hours of intervention throughout admission was completed. There were 932 hours of therapy with 934 occasions of service over a 9-month inpatient intervention period.



**Table 1: Comparison of paediatric functional assessment measure (WeeFIM)<sup>9</sup> results at admission and after 9 months of rehabilitation**

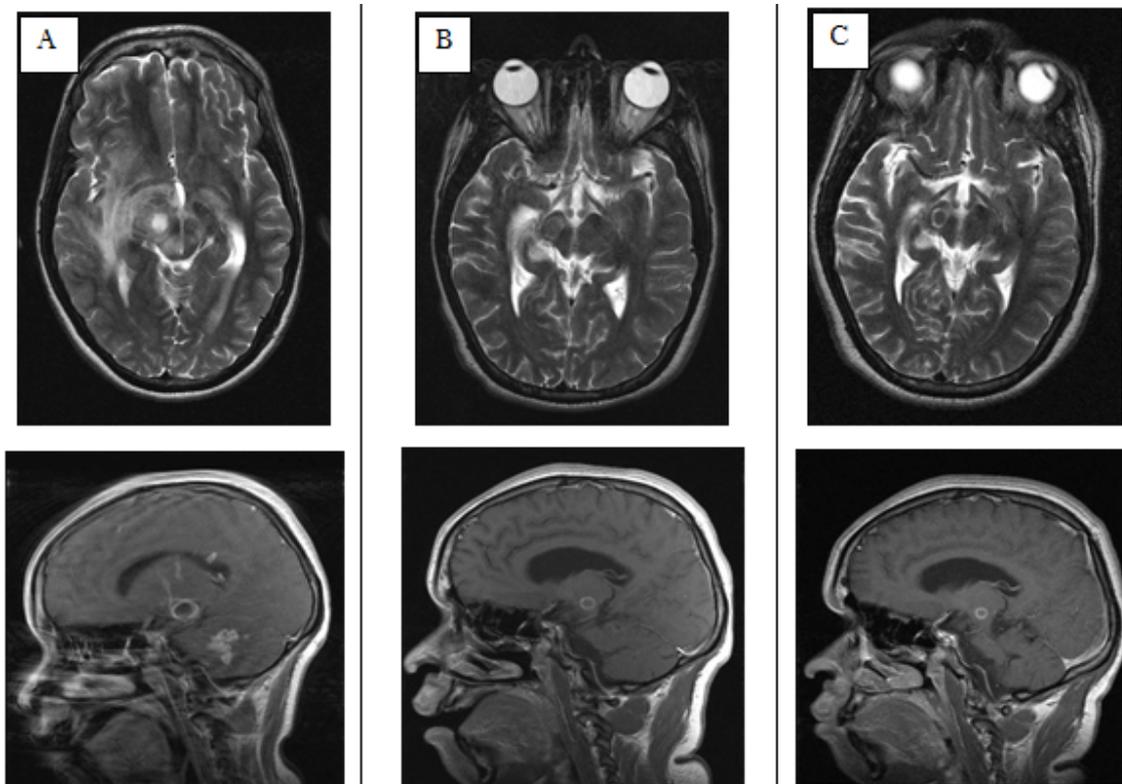
Functional independence measure (WeeFIM) domains	Initial assessment	After 9 months rehabilitation
Eating	1	6
Grooming	1	2
Bathing	1	2
Dressing – upper body	1	3
Dressing – lower body	1	1
Toileting	1	1
Bladder management	1	3
Bowel management	1	2
Transfer – chair, wheelchair	1	2
Transfer – toilet	1	2
Transfer – tub, shower	1	2
Locomotion – walk, wheelchair, crawl	1	1
Locomotion – stairs	1	1
Comprehension	1	6
Expression	1	6
Social interaction	1	5
Problem solving	1	2
Memory	1	6

Discharge planning was commenced early during his admission owing to the remote location of his home town (more than 1000 km from metropolitan services) and its limited resources (nursing outpost only). It was identified that a high level of functional independence was required in order to achieve a safe discharge home, necessitating a longer inpatient rehabilitation stay. Significant efforts were made to link with his remote community. Telehealth services, social media and video diaries facilitated connection with family, health and disability providers in his community and with local Aboriginal elders to guide the planning process.

Functional gains began to emerge and consolidate at approximately 4 months. Consistent communication was established through a combination of eye gaze, gestures, key word signs and facial expressions. Although severely dysarthric he could pronounce some single words, which progressed into simple sentences. He did not appear to have a marked cognitive deficit although recent school reports were

difficult to obtain for comparison owing to limited school attendance. He was able to access a computer and tablet device with supports and could, for the first time, type a sentence to express his wishes and feelings. He commenced standing transfers with the assistance of two trained staff at this time.

Towards the end of the inpatient rehabilitation stay, both the patient and his family indicated their preference to return home to their local community upon discharge. They acknowledged the distance from health and other support services; however, connection to country was an important component of this patient's identity and sense of wellbeing. Emphasis of intervention was then focused heavily towards carer and family training to ensure safety and the prevention of further complications such as pressure sores and regression in functional skills.



**Figure 1: Standardized magnetic resonance imaging views showing melioidosis progression. (A) 3 months: Multifocal ring-enhancing lesions scattered throughout both cerebral hemispheres. (B) 6 months: Reduction of oedema in both cerebral hemispheres. Ring-enhancing lesion within the right cerebral peduncle smaller than previously. (C) 9 months: Ring-enhancing lesions remain, with noticeable reduction in size, especially within the cerebral peduncle.**

Long-term medical management included ongoing prescription of Sinemet (levodopa-carbidopa), which markedly improved his signs of bradykinesia and tremor. He also required botulinum toxin A injections to his gastrocnemius to manage spasticity and maintain adequate ankle range of movement for weight bearing. He completed a 9-month course of co-trimoxazole.

### *Follow-up and outcomes*

The patient's functional assessment at discharge demonstrated that he had attained voluntary control of all

four limbs, and despite his truncal ataxia and coordination impairment he could perform a standing transfer with the assistance of one person. He could ambulate short distances with the assistance of two people; however, due to the ongoing deficits in his motor planning, initiation and motor control, this was of limited functional utility. His range of motion was maintained during admission with serial casting and splinting. He was able to independently drive a powered wheelchair as his primary form of independent mobility. He required full assistance from a carer for self-care tasks such as showering, dressing and toileting. Scoring of his WeeFIM<sup>9</sup> assessment was completed and he achieved 53/126,



indicating significant need for ongoing assistance in multiple domains (Table 1). The Pediatric Test of Brain Injury, an assessment of cognitive linguistic abilities including memory, attention and language<sup>10</sup>, was administered. Subtests demonstrated strong verbal comprehension skills, good vocabulary and semantic skills and the ability to communicate in sentences.

The WeeFIM<sup>9</sup> was used to document recovery throughout the admission. This was scored by a therapist trained in administering the assessment. The WeeFIM is a valid outcome measure commonly used in acquired brain injury rehabilitation settings<sup>11,12</sup> and is an adaptation of the original FIM. It can be used to document functional and cognitive status throughout admission across a seven-level ordinal scale (1 = total assistance, 7 = complete independence), providing scores across six key domains: self-care, sphincter control, transfers, locomotion, communication and social cognition<sup>9</sup>. The maximum score possible (126) is equivalent to the developmental age of a 7-year-old child<sup>11</sup>.

Table 1 shows the trajectory of recovery as determined by assessment with the WeeFIM. Initially, the patient scored 18/126, the lowest possible score, indicating the requirement for total assistance in all domains of assessment. After 9 months of intensive rehabilitation he scored 53/126, demonstrating improvement but with ongoing significant dependence.

## Lessons learned

Our aim was to document the trajectory of recovery and intensity of rehabilitation required for the patient to achieve a level of functional independence that would enable a safe discharge to a remote community. This is the first known case of paediatric neurological melioidosis in Western Australia.

The common features of neurological manifestation of melioidosis include unilateral limb weakness, mixed cerebellar and brainstem features, peripheral weakness and

flaccid paralysis, fluctuating levels of consciousness, cranial nerve palsy (unilateral VII cranial nerve), abscess formation and prominent headache and fever<sup>3,5</sup>. In a review of 12 cases of neurological melioidosis presenting to Townsville Hospital between 1995 and 2011, 90% had a fever (>38°C by digital ear thermometer), 70% had a normal GCS, 70% presented with headache and 50% presented with ataxic gait patterns<sup>6</sup>.

Known risk factors for contracting melioidosis include indigenous background, diabetes, hazardous alcohol use, chronic lung disease, chronic renal disease, rheumatic heart disease and/or congestive cardiac failure, malignancy, immunosuppressive therapy and kava use<sup>1</sup>. Currie et al reported that 52% of all their melioidosis cases were indigenous Australians<sup>1</sup>, and Edmond et al (2001) reported that the average annual incidence in the paediatric population in the Northern Territory was three times higher in the indigenous population than non-indigenous population<sup>4</sup>.

The patient in the present study is thought to have contracted melioidosis whilst swimming in a flooded water course during the northern Australian wet season. His only identified risk factor was his indigenous ethnicity although he was known to have engaged in the recreational use of solvent inhalation in the years prior to presentation. Literature suggests that infection often occurs in the paediatric population without pre-existing risk factors<sup>2</sup>. There is currently no identified link between solvent inhalation and melioidosis.

Discharge planning to remote communities in Australia can be complex and slow moving due to the distance between hospital and home, and the difficulty this poses for family and carers to attend the hospital regularly. To navigate this barrier, videoconferencing facilities (telehealth) were used. The use of telehealth in Australian paediatric services appears limited; however, evidence of its successful implementation can be found in the literature<sup>13</sup>. In the present case, telehealth enabled regular communication between the treating team, regional healthcare staff and the family. This facilitated the engagement of the community in the discharge planning process and encouraged a practical problem-solving



approach. As was the case for Jury et al<sup>13</sup> in their telehealth trial, such a process helped our team to develop a strong relationship with the family, community and regional teams, allowing successful transition to a nearby regional centre whilst environmental modifications were completed at home.

This is the first documented rehabilitation case report of a paediatric patient with severe long-term neurological sequelae from melioidosis. It paves the way for greater understanding of the trajectory of recovery following onset of infection and highlights the needs for long-term rehabilitation input in order to reach the highest level of independent function possible.

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