Benign acute childhood myositis
Laboratory and clinical features

M.T. Mackay, MB BS, FRACP; A.J. Kornberg, MB BS(Hons), FRACP; L.K. Shield, BSc(Med), MB BS, FRACP; and X. Dennett, BSci, PhD

**Article abstract—**Background: Benign acute myositis of childhood is a disorder of midchildhood, typically affecting boys. Symptoms include calf pain and difficulty walking after a viral illness. There is an epidemiologic association with influenza. **Objectives:** To describe the clinical and laboratory features of benign acute myositis. **Results:** Thirty-eight children (32 boys, 6 girls) were seen with 41 episodes of myositis between 1978 and 1997. Two were siblings and three had recurrent episodes. Mean age at onset of symptoms was 8.1 years. Children remained ambulant during 33 of 41 episodes. Two characteristic gaits were noted: toe-walking in 13, with a wide-based stiff-legged gait in another 7. Muscle tenderness was isolated to the gastrocnemius–soleus muscles in 82% of episodes. Recovery occurred within 1 week. Creatine kinase levels were elevated during all episodes. Viral studies were positive in 10 of 24 episodes, 5 because of influenza B. **Conclusion:** Benign acute myositis is a syndrome of midchildhood that can be differentiated from more serious causes of walking difficulty by the presence of calf tenderness, normal power, intact tendon reflexes, and elevated creatine kinase. The gait patterns noted may minimize power generation of the calf muscles by splinting the ankles. Onset in childhood may reflect an age-related response to viral infection, and occurrence primarily in boys may reflect a genetic predisposition or an as-yet unknown metabolic defect. **Key words:** Myositis—Influenza—Guillain–Barré syndrome—Viral myositis.

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Benign acute childhood myositis (BACM) was first described in 1957 by Lundberg under the name of “Myalgia Cruris Epidemica.” It is characterized by the sudden onset of calf pain and refusal or difficulty to walk, often after a prodromal viral upper respiratory illness. There often is leukopenia and viral agents, particularly influenza B, that have been implicated in many cases. The serum creatine kinase (CK) usually is elevated. Boys are affected more commonly, and it is followed by rapid recovery usually within a week. It is rarely reported in adults.

We report 38 children with 41 episodes of BACM occurring between 1978 and 1997. We review the literature and discuss the epidemiologic correlates, in particular the association with influenza infection.

**Illustrative cases.** Case 1. A previously well 7-year-old boy was seen in June 1997 with a 5-day history of fever, cough, and lethargy. On the day of admission, he had woken at 3:00 AM reporting calf pain and later that morning had difficulty walking. On examination, he was febrile (38.9 °C) and was tender in both calves with pain on passive ankle dorsiflexion. He walked on his toes. Tone, power, tendon reflexes, and sensation were normal in lower and upper limbs.

CK was 4762 U/L (normal range, 40–240 U/L), erythrocyte sedimentation rate (ESR) was 12 mm/h (normal, <6 mm/h), and full blood count showed leukopenia (3.0 × 10^9 [normal range, 5.5–15.5 × 10^9]) with neutropenia (1.4 × 10^9 [normal range, 1.5–8.5 × 10^9]) and lymphopenia (1.2 × 10^9 [normal range, 2.0–8.0 × 10^9]) on differential. Parainfluenza 3 was isolated on rapid-enhanced culture immunofluorescence from nasopharyngeal aspirate (NPA). Within 24 hours, he was able to walk better and had recovered completely when reviewed 6 days later.

Case 2. A 9-year-old boy was admitted in September 1988 with a 4-day history of fever, cough, lethargy, and a...
1-day history of painful calves and difficulty walking. On admission, his temperature was 38.5 °C, and his gait was wide-based and high stepping. Both gastrocnemius–soleus muscles were tender, but power in the lower limbs was normal. The right ankle jerk was thought to be absent. Apart from limitation of left lateral gaze consistent with Duane syndrome, the rest of the neurologic examination was normal.

Urinalysis on dipstick was negative for blood. CK was 1254 U/L (normal range, 40–240 U/L). Full blood examination showed leukopenia (3.0 × 10⁹) with a normal hemoglobin and platelet count. ESR was 10 mm/h. Serologic analysis for influenza A, influenza B, and toxoplasmosis was negative by complement fixation.

Mycoplasma pneumoniae immunoglobulin M was equivocal and immunoglobulin G was positive. He walked better the next day and had made a complete recovery within 3 days.

After a 4-day history of fever and lethargy in September 1989, he had recurrence of symptoms. He had woken at 5 AM on the day of admission with painful legs and was unable to stand or walk. On examination later that day, his temperature was 38.2 °C, and he had tenderness of the thighs and gastrocnemius–soleus muscles. He walked (with assistance) with the trunk flexed at the hips and both knees extended. Tone, power, and tendon reflexes were normal in the lower limbs.

The leukocyte count was 3.0 × 10⁹ with neutropenia (0.9 × 10⁹) and a normal lymphocyte count. ESR was 2 mm/h. Urea and electrolytes were normal. Serology was negative for influenza A and B, M. pneumoniae, and toxoplasmosis. Viral throat swab isolated herpes simplex. Convalescent serum for herpes serology was not obtained.

Muscle biopsy of the left calf was performed, as there were concerns about an underlying metabolic myopathy. This showed fibers undergoing active degeneration with necrosis, myophagia, and ghost fiber formation in most. A mild increase in fat was seen in some fibers (figure). Enzyme histochemistry was normal, with a normal checkerboard pattern and normal staining for glycogen, myophosphorylase, and phosphofructokinase. The findings were consistent with acute segmental degeneration, consistent with a monophasic myositis. Muscle carnitine palmitoyl transferase activity was normal. Recovery was complete. Convalescent CK 3 weeks later was 149 U/L (normal range, 40–240 U/L).

Case 3. The younger brother of the Case 2 patient was examined in August 1991, aged 11, after a 3-week history of cough, fever, and lethargy. Three days before admission, he developed calf and quadriceps pain associated with difficulty walking. On examination, he was febrile (37.8 °C) and had tenderness of the gastrocnemius–soleus muscles with restriction of passive dorsiflexion due to pain. He walked on his toes. Neurologic examination was otherwise normal.

Acute CK was 11,700 U/L (normal range, 40–240 U/L). ESR was 5 mm/h, and there was leukopenia (3.8 × 10⁹) with lymphopenia (1.4 × 10⁹). Adenovirus and parainfluenza were isolated on NPA. Symptoms resolved within 5 days. Convalescent CK 1 month later was 82 U/L.

Materials and methods. Between August 1978 and October 1997, 34 children with BACM were seen at the Royal Children’s Hospital (RCH), Melbourne, 1 child at a Melbourne metropolitan hospital, and 3 children at a hospital in regional Victoria. Children seen before 1997 were ascertained by searching the RCH Department of Neurology database, and each child’s medical record was reviewed by the authors. In 1997, all children with symptoms suggestive of BACM and seen at the RCH were referred to the neurology department and examined by one of the authors. Follow-up was by review in the emergency or neurology department or by telephone interview. In the four children seen at other centers, clinical and laboratory data were obtained by correspondence with the children’s pediatricians. Inclusion criteria were acute onset of calf pain and difficulty walking. For children to be included in this cohort, adequate documentation of general and neurologic examinations was required and the illness needed to be self-limited. Laboratory data were not required for inclusion.

Six children with a possible diagnosis of BACM were not included. Two children were seen in the neurology clinic because of longstanding toe-walking. They had episodes 2 and 6 months previously that were consistent with BACM, but because there was inadequate documentation of the episodes, they were excluded from analysis. One
child with a history of spastic cerebral palsy and intellectual disability was evaluated 4 months after onset with persistent pain, difficulty walking, and elevated CK level. She was excluded because of persistent symptoms, and her intellectual disability did not allow adequate evaluation. A further three children were excluded because of inadequate documentation of the history, general, and neurologic examination. These children had recovered quickly after onset.

Over the course of the study period, the normal reference range for CK changed. Before August 1980, the normal reference range was <80 U/L; afterward, the normal reference range was 40 to 240 U/L.

Viral studies included throat swab for viral culture and nasopharyngeal aspirate, which was examined by direct immunofluorescence and rapid enhanced tissue culture for influenza A and B; parainfluenza 1, 2, and 3; adenovirus, respiratory syncytial virus (RSV), and cytomegalovirus. Viral serology was performed using hemagglutinating and complement fixing antibodies against influenza A and B, adenovirus, and *M. pneumoniae*.

**Results.** **Demographic data.** Thirty-eight children with BACM were seen; 3 of these children had a second episode. Mean age at onset of symptoms was 8.1 years with a range of 3 to 13 years. Thirty-two (84%) were boys and 6 (16%) were girls. Two children were siblings, one of whom had a recurrence. There were two outbreaks of BACM episodes in 1978 (8 episodes), 1991 (8 episodes), and 1997 (15 episodes). All cases in 1978 and 1991 and 9 of 15 cases in 1997 occurred in the winter months. Overall, 27 cases occurred in the winter, 7 in the fall, 6 in the spring, and only 1 in the summer months.

**Clinical findings.** Prodromal symptoms included fever (80%), cough (59%), headache (37%), rhinorrhea (20%), sore throat (17%), and vomiting (17%). Other prodromal symptoms included abdominal pain, diarrhea, neck stiffness, earache, lethargy, malaise, and irritability. The average duration of the prodrome was 5 days, with a range of 2 days to 3 weeks. In 13 BACM episodes (32%), onset of myositis began between 1 and 4 days after resolution of the prodromal symptoms. In the other 68%, onset was concurrent with the prodrome.

A provisional diagnosis of viral myositis was made during 5 of the 21 BACM episodes in children initially seen by general practitioners. Guillain–Barré syndrome was the most common alternative diagnosis (six episodes) with other diagnoses including polio (two), transverse myelitis (one), acute cerebellar ataxia (one), viral illness (one), and conversion disorder (one).

At the time of onset of 36 episodes of BACM, 12 children (33%) were afebrile, 15 (42%) had a temperature <38 °C, and 9 (25%) had a temperature >38 °C.

The gait was abnormal for all episodes of BACM. Eight children (19%) were unable to walk and in those who could walk, two characteristic patterns were observed. Thirteen (32%) children walked on their toes and resisted attempts at passive ankle dorsiflexion. Another seven (17%) walked with a wide-based, stiff-legged gait. The trunk was flexed forward on the hips, the legs were externally rotated at the hips, and the knees were locked in extension. For the remaining 13 (32%) episodes, gait was described as bizarre, unusual, wide-based, stumbling, stiff-legged, or antalgic.

**Laboratory studies.** Urinalysis was abnormal in 3 of 14 tested. One child with a history of nephrotic syndrome had 0.3 g/L of protein on dipstick. Two other children were positive for blood on dipstick, but urine was macroscopically normal in color. In the second child, microscopy revealed 4 × 10⁶ erythrocytes/mm³. CK level was 248 U/L (normal range, 40–240 U/L). In the third child, there were 8 × 10⁶ erythrocytes/mm³, but urine was negative for myoglobin. CK level was 7029 U/L.

CK estimation was performed in the acute phase of 37 BACM episodes. This ranged from 248 U/L (just above the normal range of 40–240 U/L) to 14,154 U/L (59 times normal) with a mean of 3271 U/L (14 times normal). In the child with a CK level of 14,154, urinalysis was not performed. In the other child with a CK level above 10,000, urine was normal. No child tested had a normal CK level in the acute phase. Convalescent CK performed after 12 episodes was normal in all cases. ESR measured during 19 episodes was normal (<6 mm/h) in 10 children (53%), <15 mm/h in 7 (37%) children, and 20 mm/h and 25 mm/h, respectively, in 2 children. Leukopenia was noted in 17 (68%) of 25 episodes, and of these, 11 were neutropenic, 2 lymphopenic, and both differentials were decreased in 4.

Viral studies were positive in 10 (42%) of 24 episodes. Influenza B was isolated in five episodes, influenza A in one episode, parainfluenza in two episodes, both adenovirus and influenza B in one episode, and herpes simplex in one episode. In 1978, of six cases tested, all three isolates were influenza B, whereas in 1989, of the three cases tested, one isolate was parainfluenza and another was herpes simplex. In 1991, of the two cases tested, influenza B and adenovirus were identified in the one isolate. In 1997, of the 15 cases tested, 2 were influenza B, 1 was influenza A, and 1 was parainfluenza type-3. Muscle biopsy was performed on one child with recurrent myositis (see illustrative Case 2).

**Epidemiology of influenza.** Laboratory data for most confirmed cases of influenza A and B are recorded at two laboratories in Victoria: the Victorian Infectious Diseases Reference Laboratory (VIDRL) and the Department of Virology, RCH. Records of confirmed cases have been kept from 1984 onward at VIDRL and from 1985 onward at the RCH.

The data show that influenza A epidemics occur each year, whereas influenza B cocirculates with influenza A in odd-numbered years. Influenza A was particularly prevalent in the years 1988, 1992, and 1997, and influenza B was prevalent in the years 1985 and 1997.
Most of those who could walk fitted into one of two gait patterns. In our series, 84% of patients were boys compared to 64% in earlier studies. Some authors have noted onset of calf pain after strenuous exercise the previous day and postulated that the increased incidence in boys may be related to greater levels of activity compared with that of girls.6 During influenza epidemics, only a small proportion of children have BACM develop. This, combined with the disproportionate number of boys; recurrent episodes in some children1,4,5; and involvement of siblings, raises the possibility that BACM occurs in genetically susceptible individuals with a metabolic defect of muscle provoked by a viral trigger. In our second illustrative case, biopsy failed to reveal any metabolic defect.

The mean age at onset of symptoms in our series was 8.1 years, which is similar to that recorded in previous studies of 9.2 years.1,3,4,6-9 Apart from the report by Lundberg4 of four adults, BACM has not been reported in adults. One author has described a 19-year-old woman with postviral myositis in whom virus-like particles were identified by electron microscopy on muscle biopsy, but it is unclear whether the severe myalgia primarily affected the calves.10

The mean duration of prodromal symptoms in our study was 5 days compared to 3½ days in previous studies.1,3,5 Several authors have noted onset of calf pain after a period of rest, often on waking in the morning, and with the maximum pain coinciding with return of the temperature to normal,2,4 whereas myalgia associated with influenza in adults occurs concurrently with the viral symptoms. In our series, 13 (32%) of BACM episodes occurred after resolution of prodromal symptoms.

Myalgia primarily affects the gastrocnemius and soleus muscles. In 82% of our children and 91% of cases in previous series, the calves were involved in isolation, but other muscle groups sometimes involved include the thighs, back, neck, and proximal arm muscles.1,6 Most of those who could walk fitted into one of two gait abnormalities, an observation not described previously in detail. Some walked with a wide-based, stiff-legged gait with the trunk flexed on the hips and knees extended. Others toe-walked. Both groups, by splinting the ankles, avoided stretching of the calves and reduced power generation in the gastrocnemius–soleus complex. This is further supported by the observation that passive ankle dorsiflexion increases pain.

Resolution is rapid, usually within 1 week, although some children in the literature took up to 7 weeks.11 Elevated CK is characteristic and was identified in all our children tested and in 94% of those reported in the literature.2,4,11 This typically returns to normal within 1 month. Leukopenia is commonly seen (68% in our series compared to 73% in the literature). ESR is elevated in fewer than half of the cases and usually is not >20 mm/h.

BACM can occur sporadically or in epidemics. Since the first report by Middleton et al.,2 several authors have confirmed the association with influenza B. Other viruses including influenza A, parainfluenza, adenovirus, Coxsackie, and M. pneumoniae have been isolated less frequently.2,5,7 The isolation of herpes simplex in one of our children is of uncertain significance as serologic studies were not performed.

Recurrence of BACM has been reported previously in only seven cases.1,2,4,5 In our series, there were three recurrent cases. This observation, along with the lack of reports of BACM in adults, led Dietzman et al.3 to propose that the occurrence of BACM in midchildhood reflected initial exposure to influenza. This was based on their observation that all previously reported patients with BACM had negative acute serology for influenza. Ruff and Secrist5 showed, in a series of 33 children with BACM, that all had negative acute sera for either influenza A or B followed by a positive titer on convalescent serum. There were two children in that series who had recurrent episodes, and they showed the first episode was caused by influenza B and the second caused by influenza A, further supporting this hypothesis. These two children had a further episode of influenza B and did not have BACM develop.

In our series, 42% of those tested had positive viral isolates or serology with 50% of these cases caused by influenza B. In the previous larger studies, the range of positive viral studies was between 65% and 100%, and influenza B was isolated in most of these.2,4,12 Although influenza B was only isolated in two of the four positive viral studies in 1997, the large number of cases of BACM compared with that of previous years would fit with the 1997 epidemiologic data from VIDRL and RCH that showed influenza B was more prevalent in the Victorian community in 1997.

It is important to differentiate BACM from more serious causes of refusal to walk or limb pain. Guillain–Barré syndrome was considered in a number of our children, but the normal power and reflexes in most cases combined with elevated CK argue against this diagnosis. Although entries in the medical records of some children suggested that there was...
muscle weakness, from our observations, we are doubtful that weakness was actually present. It is most likely that failure to generate power because of pain was attributed to weakness.

The pathogenesis of BACM remains unclear. Muscle biopsies have been performed in 19 cases reported in the literature, and pathologic changes identified included nonspecific degenerative changes, focal muscle fiber vacuolation, and extensive muscle necrosis.\(^1,4,5,7,8\) In one child, muscle necrosis was found in a deltoid biopsy specimen without clinical involvement of that muscle, suggesting the myositis is more widespread than expected from clinical examination.\(^5\) We believe that the term myositis is justified as there was evidence of an inflammatory component to this disorder in the patient who underwent biopsy in our series and in some muscle biopsies described in the literature. A significant number of muscle biopsy specimens have not shown an inflammatory component; however, pathology may be lacking because the process could be patchy. The marked elevation of CK suggests muscle fiber destruction in keeping with a myositic component.

Influenza can produce a spectrum of muscle disorders including myalgia, BACM, and acute generalized myositis.\(^13\) Although influenza and other viruses have rarely been isolated from muscle in other forms of viral myositis,\(^13,14\) there is only one report in the literature of the results of a muscle biopsy specimen being positive for influenza B in BACM.\(^4\) Therefore, uncertainty remains over whether the myositis is caused by direct viral invasion or by immune-mediated mechanisms.\(^5\) The occurrence of BACM in midchildhood may reflect an age-related response of muscle to viral infection.

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