

Letters to the Editor

Salmonella arizonae Infection from Snake Bite

To the Editor: Snakes, lizards, terrapins and other reptiles are the main natural reservoir of *Salmonella arizonae* (*S. arizonae*).¹ Several investigators have reported the fact that snakes and other reptiles harbor and transmit *S. arizonae* to humans.²⁻⁵ Therefore, whenever and wherever human infection due to this organism occurs, it should trigger suspicion of a possible connection with reptiles as well as poultry and egg products. In areas of Saudi Arabia where snakebite is common, deliberate attempts should be made to exclude secondary bacterial infection due to *S. arizonae*, which could be fatal. Our report highlights a case of *S. arizonae* wound infection in a Saudi boy bitten by a snake.

Case Report

A 10-year-old Saudi boy presented at our Emergency Department in November 2000 with a history of snakebite on the dorsum right big toe, having been referred from a primary health care center (PHCC), where he received polyvalent anti-snake venom (equine serum) and a booster dose of tetanus toxoid. He did not develop any systemic manifestation of the complication of snakebite, but subsequently had abscess at the site of the bite over the big toe, for which he was referred to the Asir Central Hospital in Abha.

On admission, the patient was found to be healthy-looking and afebrile, with an abscess over the dorsum of the first metacarpophalangeal joint of the right big toe. At the center of the abscess was a black penetrating wound mark (caused by the fangs of the snake) surrounded by an induration and edema. The movement of the joint was limited, and a diagnosis of septic arthritis of the metacarpophalangeal joint with overlying abscess was made.

After preliminary investigations, the abscess was drained and the pus was sent for Gram's stain, culture and sensitivity. The patient was admitted and put on intravenous cefuroxime and amikacin 6 hourly pending culture results. X-ray of the affected toe revealed no bony involvement. Results of the other investigations carried out showed PT of 19 (control 17), APTT 34 (control 30), ESR 22 mm/hour, urea 41, creatinine 0.7, RBS 96, Na 131, and K of 3.4.

Pus from the infected toe yielded pure growth of *S. arizonae* which was sensitive to ampicillin, augmentin, cefuroxime, cefotaxime, ceftriaxone, chloramphenicol, gentamicin, amikacin, co-trimoxazole, and imipenem. *S. arizonae* was identified using API 20E strip (BioMerieux Vitek Inc., Missouri, USA). After the laboratory report, the patient was reviewed and found to be stable and in

satisfactory condition. He was discharged home to continue the dressing of his wound at the PHCC. He defaulted on follow-up and was presumed cured.

Discussion

Reptiles are the main natural reservoir of *S. arizonae* but humans, poultry and other animals have also developed infection from this organism.¹ Nine out of ten reptiles shed salmonellae in their feces, and attempts at their elimination and treatment have largely failed. In the United Kingdom, the Chief Medical Officer of the Department of Health has warned that owners of snakes, lizards, terrapins and other reptiles are at the risk of contracting salmonella from their pets.⁶ In a recent report, the Public Health Laboratory Services showed an increase in salmonella cases associated with exotic pets in children and infants. The report noted that in the previous two years (1999 and 2000), 13 people, including a three-week-old baby who died, had contracted salmonella from pet reptiles.⁶

Reports of *S. arizonae* infection in cases of snakebite² and in cancer patients and HIV-positive patients following consumption of snake powder capsules as a so-called "folk remedy" have been reported by many investigators.⁷⁻¹⁰ The clinical spectrum of *S. arizonae* infections varies and may include benign gastroenteritis, enteric fever and septicemia, localized infection in diverse organ sites such as the brain, bone, liver, lung, joints and gall bladder, which may or may not be a sequel to septicemia. Well-documented cases of osteomyelitis caused by *S. arizonae* have been reported.¹¹ However, our patient did not develop osteomyelitis of the affected toe, possibly because the infection was detected early and managed appropriately. We wish to draw the attention of clinicians and laboratory staff to the possible presence of this organism in cases of wounds caused by bites of reptiles, especially snakes, both wild and domestic.

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Osteopenia and Osteoporosis in Two Children with Graves' Disease

To the Editor: The adverse effects of long-standing untreated hyperthyroidism on the bone were first described in 1891 by von Reckling Hausen in a 29-year-old woman who died after five years of hyperthyroidism. The changes described were predominantly those of bone destruction, with increased osteoclastic and osteoblastic activity, especially in cortical bones, causing high turnover osteoporosis and fractures. These changes have been described extensively in adults, but data in children are lacking. We report two cases of Graves' disease in children who had significant osteopenia and osteoporosis at diagnosis.

Case 1

The patient was an 11-year-old Libyan girl who presented with low back pain after sustaining a fall on her back a day earlier. For the following two weeks, she complained of generalized weakness, shakiness of her hands and a protrusion of her left eye. There had also been a noticeable weight loss over the previous few months. Her past medical history was significant for congenital strabismus, for which she had repeated surgeries that had resulted in significant proptosis of the right eye. She had had speech delay since early childhood, which worsened with the recent tremors and weakness. On physical examination, she looked severely underweight (19 kg, below the 3rd centile), with a heart rate of 130/min., and blood pressure of 122/65 mm Hg. Her hands were warm, with obvious tremors bilaterally, and the eyes were proptotic, especially on the right side. She had significant kyphosis and tenderness on the thoracolumbar spine. A clinical diagnosis of Graves' disease was confirmed by biochemical findings of FT₄ of 69.1 pmol/L and TSH of

less than 0.03 mU/L. Radiological examination of her spine revealed compression fracture of the T₆ and T₁₀, as well as

generalized severe osteopenia. Dual energy x-ray absorptiometry (DEXA) scan showed a significant loss of bone marrow density (BMD) in the lumbar spine as well as the left hip. Other results included: PTH 70 ng/L (n=10-65), Ca 2.29 mmol/L (2.25-2.62), PO₄ 1.77 mmol/L (1.07-1.71), alkaline phosphatase (ALP) 279 IU/L (160-500), and 25-HO vitamin D of 52 pmol/L (N=25-90). She was started on methimazole 20 mg daily with Ca 300 mg/kg/d and Vitamin D supplements (1.25 dihydroxy-cholecalciferol, 0.25 mcg/d), and was encouraged to undertake active exercise. She showed significant clinical improvement in terms of her weakness and tremors. Compliance was a major problem. Her Ca, phosphorus and alkaline phosphatase were repeated once and were all normal. She was subsequently lost to follow-up for more than a year. A repeat DEXA scan was not done.

Case 2

A 14-year-old boy presented with neck swelling, polyphagia, weight loss, increased sweating, nervousness and insomnia for a period of three weeks. He had previously been healthy and was not on any medications. His normal diet was rich in calcium (dairy products and vegetables), and he was active and athletic. Physical examination revealed an anxious, thinly built boy (37.7 kg, below the 3rd centile). He had exophthalmus, lid lag and retraction. The thyroid was diffusely enlarged with positive bruit. His hands were warm, sweaty and tremulous. His heart rate was 110/min., and blood pressure was 140/80. He was diagnosed with Graves' disease and was started on methimazole 30 mg daily, as well as propranolol. Thyroid function test showed a thyroxine level of 194 nmol/L, free thyroxine of 76.1 pmol/L and TSH of less than 0.01 mU/L. The thyroid scan was consistent with Graves' disease. Initial DEXA scan showed significant osteopenia with calcium levels of 2.44 mmol/L, ALP of 467 U/L and phosphate of 1.8 mmol/L. He was continued on methimazole for two years, during which his thyroid function was monitored regularly. He went into remission after 18 months. The methimazole dose was reduced, and finally stopped after six months. His Ca, phosphorus and alkaline phosphatase were repeated at remission and were all normal. He is currently seven months into remission, and an assessment of his BMD has shown no evidence of osteopenia.

Discussion

Our patients clearly demonstrate that children with Graves' disease can have a significant reduction in their BMD at diagnosis. Although this has been well established in adults,^{1,2} only a few studies have been reported in children.^{3,4} Our first patient had decreased BMD in the

lumbar spine and the hip, although for the hip there was no age-matched control. The second patient had a low BMD in the lumbar spine as well, but there was no age-matched control either. These results are in accordance with earlier reports in adults.^{5,6}

Lucidarme et al.³ have demonstrated that children with Graves' disease have significant reductions in BMD at presentation, and that femoral and lumbar spine BMD are significantly lower than expected for age and sex. Leger et al.⁴ reported the case of two children aged two years who presented with Graves' disease and severe bone demineralization. One had spontaneous fractures and collapse in the lumbar vertebrae. The second had complete resolution of his osteopenia after attainment of euthyroidism. This condition has been reported in adults,^{7,8} although the degree of recovery of osteopenia has been variable. Lucidarme et al.³ also reported significant gains in femoral ($P=0.001$) and lumbar spine ($P=0.02$) BMD during treatment, and none of his 11 patients showed osteopenia. Our first patient had normal Ca, ALP, 25 OH Vit D and PTH, and the second had normal Ca and ALP. This is in agreement with the study by Leger et al.⁴ Siddiqi et al.⁹ reported a rise in the ALP coinciding with euthyroidism at 4-8 weeks, and he attributed that to continuing bone formation. The mean serum OC level was increased in Leger's study in the children with hyperthyroidism compared with that in the control group, however, no correlation was found between OC levels and the individual BMD and the thyroid hormone levels. Similar findings were reported in adults.^{1,8} The effect of Ca, Vit D supplementation and physical activity needs to be evaluated.

In conclusion, our patients, as well as those recently reported, show that Graves' disease can be associated with significant bone demineralization at presentation. The changes are reversible as euthyroidism is achieved with treatment. This was clear from the literature,³ as well as from our second case. Whether we need to do a BMD test in all children presenting with Graves' disease is still to be decided since the effect is reversible, and finding age-matched controls for BMD in the young age group has not yet been done.

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Primary Pulmonary Alveolar Proteinosis: A Case Report and a Review of the Literature

To the Editor: We read with interest the article of Wali et al.¹ We have also encountered two similar cases of pulmonary alveolar proteinosis (PAP) with widespread appearance of ground glass opacities (Figures 1 and 2), involving two adults aged 43 years. Diagnoses were made with bronchial alveolar lavage fluid. In one case, the diagnosis was difficult because of pulmonary hemosiderosis. The cases have been followed for several years. Garcia Rio et al.² have published a series of six cases. In one patient, pulmonary tuberculosis appeared three months after the diagnosis of PAP. Both diseases disappeared with anti-tuberculous treatment.

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FIGURE 1. Extensive bilateral lesions (Pr Anthoine).

FIGURE 2. Predominantly right-sided lesions (Pr Bernadac).

Reply

To the Editor: I would like to thank Dr. Hoeffel and colleagues for their comments and interest in the said article. I wish to comment on the diagnosis of alveolar proteinosis by bronchial lavage cytology only and on the response of Dr. Hoeffel's patient to anti-tuberculosis medication only. As mentioned in our review, bronchoalveolar lavage (BAL) cytology alone has been shown to be a reliable tool in confirming the diagnosis of alveolar proteinosis in the right clinical setting.^{1,2} This is again in keeping with Dr. Hoeffel's cases. More recently, the diagnosis of pulmonary alveolar proteinosis (PAP) has been performed by the demonstration of autoantibodies against GM-CSF, although this appears to be highly sensitive (further confirmation studies are needed) in cases of idiopathic PAP, but less sensitive for secondary PAP.³ To date, the only successful treatment for PAP is by a whole lung lavage regardless of whether it is a primary or secondary disorder.¹ As is known, the natural history of the disease is variable, which makes the evaluation of the efficacy of any specific treatment difficult. Therefore, it is not clear whether the reason for the recovery of Dr. Hoeffel's patient was spontaneous or related to the response to anti-tuberculous drugs. On the other hand, patients with PAP are susceptible to chest infection, and hence the appearance of pulmonary tuberculosis may represent a superadded infection, rather than a triggering factor for alveolar proteinosis.

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Fatal Child Abuse in Two Children of a Family: The Alleged Role of Polygamy

To the Editor: We would like to thank Dr. Elkerdani et al.¹ and Dr. Maziak² for tackling such a sensitive medicolegal

issue. Child abuse is the maltreatment of a child by an adult in a way which is unacceptable in a given culture at a given time.³ Child abuse is subdivided into physical, sexual, and emotional abuse and child neglect.⁴ This sociomedicolegal condition is very common in the West. In the US, an estimate of 1.6%-14% has been reported, out of which 4000 children die annually.⁵⁻⁷ In the UK, 4% of children less than 12 years suffer from child abuse, 1 in 10,000 dies, and the incidence is increasing.³ We are quoting these records just for comparison with the incidence of child abuse in Saudi Arabia, where it is a newly emerging or a newly identified entity.⁸ Only a few case reports are available in the Saudi literature,⁹⁻¹³ and the total number of cases is very low.¹¹

The letter by Maziak² commenting on the reports by Elkerdani et al.¹ and Al-Ayed¹¹ focused on the social and legal links of child abuse, especially the role of polygamy as a risk factor. We have some reservations on this point. First, we agree with the author that anecdotal reports do not fit in scientific documentation, and that the conclusions based on them are erroneous and misleading. Unfortunately, he did so in considering polygamy as a risk factor for child abuse. A few case reports of child abuse in a country where polygamy is common is statistically insignificant. To counter this, one may quote other anecdotal reports where polygamy has been shown to save some compromised children from a psychologically unstable mother or from poverty-related child abuse.⁷ Shall we conclude then that polygamy prevents child abuse? Certainly not. However, the fact that child abuse is rare in Saudi Arabia, where polygamy is common, may itself indicate a reverse proof against this claim. Yet, a comparative study between the prevalence of child abuse in polygamous and monogamous families is required after fixing the other etiological factors in both research groups. For example, in the report of Elkerdani et al.,¹ polygamy was not possibly the only acting factor to hang the case on. There was the mother herself who was quite young (16 years old), the large difference in age between the mother and the husband, and the illiteracy of both parents. Also there was no elucidation of the mental and psychological status of the mother. It is hard not to attribute some criminal intention and aggression from a natural mother, even if she is wife No. 10, against her defenseless infants without some psychotic defect underlying it. We cannot blame one likely factor in the presence of other factors if we are neutral and scientific. Second, in a rough review of 23 Saudi cases of child abuse,^{11,13} polygamy was found in two families only, noting that some of these children belonged to unknown mothers. Third, child abuse is a multifactorial disorder.¹⁴ A review of the literature shows that polygamy has not been reported as a risk factor or predisposing factor for child abuse.³ On the contrary, it has been shown that most child abuse cases have been perpetrated by a family member, usually a parent or the mother's boyfriend.¹⁵ There is no available study on the effect of polygamy. Fourth, the author may have been

influenced by the Western experience of polygamy, which is quite different from what happens in Saudi Arabia. Polygamy or even “multi-polygamy” is widely practiced in the West, but because it is illegal, it is practiced in the dark, with no responsibility towards the child that comes out of that relationship. They rather try to get rid of the undesired pregnancy. So, child abuse there might have a strong link with such illegal polygamy, especially in a situation where the so-called child protection agencies are themselves ineffective. Thus, the legality and official acceptance change completely the prognosis of polygamy and the children born out of it. Fifth, any legal rule or official regulation, including polygamy, may be abused when put in practice. This should be dealt with separately and accordingly without undermining the rule itself. Sixth, even if we propose child abuse as an adverse side effect of polygamy, it should be weighed against the positive social effects and benefits of polygamy, which are beyond the scope of this letter. Seventh, it is completely unscientific to generalize the suggestion of child abuse on all Middle Eastern countries when the author is commenting on child abuse in Saudi Arabia, especially when polygamy is not common in other Middle Eastern countries.

Citing polygamy as a cause of child abuse is unscientific without statistical studies. We should differentiate between legal polygamy, which is practiced in Saudi Arabia to serve special social conditions, and the illegal polygamy widely practiced in the West which may be related to child abuse, and which places no responsibility towards the resulting offspring. The rare occurrence of child abuse in Saudi Arabia, where polygamy is common, may help remove any suspected relationship between them. A rough review of 23 cases of child abuse in Saudi Arabia showed that polygamy was found in only two cases.^{11,13}

NB: We are commenting on the issue on principle only. None of the authors of this article practices polygamy, so the personal factor is excluded.

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Reply

To the Editor: In response to the letter by Dr. Shelleh et al., I start by repeating the part of my published letter that initiated the current debate. I said “it is astonishing that in the two reports, the father was married to two women, and the involved mothers were the second wives, yet polygamy was not even mentioned as a *possible* cause of trouble in both cases.” I think it is clear to the reader that I am not stating that polygamy is a risk factor, let alone *the* risk factor for child abuse, as Dr. Shelleh et al. accuse me of doing. Thus, a simple, correct reading of what I have written renders their rebuttal out of context and unnecessary. Any factor surrounding the case of interest, as I have clearly stated in my letter, is a possible attribute to the case until it is proven otherwise. The acknowledgement by Dr. Shelleh et al. that the relationship between polygamy and child abuse needs to be studied is a clear support of my view that this factor, along with others, should receive due consideration. The fact that only a few cases of child abuse have been reported in Saudi Arabia compared to the large numbers in Western countries, as presented by Dr. Shelleh et al., does not mean that it is rare in Saudi Arabia compared to the West. Most developed countries have systematic tracking and surveillance systems for various public health problems, including child abuse. From the available medical literature, I am not aware of the presence of national tracking and surveillance systems for child abuse

in Saudi Arabia, which means that the assumption that it is rare compared to the West is at least premature.

In the response of Dr. Shelleh et al., they repeatedly refer to what is scientific and unscientific, of course accusing me of being unscientific and deprived of neutrality. I am just wondering where science is in their prejudiced, stereotypic description of the West as the land of shattered families, where sexual promiscuity is practiced in the darkness. And where is science in their classification of legal polygamy as practiced in Saudi Arabia vs. the illegal polygamy practiced in the West, which is based solely on their personal opinion, and with complete disregard to others' legal and cultural specificities? In fact, in their rebuttal Dr. Shelleh et al. go as far as to object even to my consideration of polygamy as a risk factor, which makes me wonder if all this is because I have trespassed on a taboo. For very mysterious reasons as well, they object to my plea for the establishment of child protecting agencies in the whole Middle East to deal with cases of child abuse, which was advanced in the context of providing legal bodies capable of taking active measures in cases of child abuse.

In their rebuttal, Shelleh et al. presented arguments in favor of polygamy having nothing to do with child abuse, and based on the fact that of the 23 cases of reported child abuse in Saudi Arabia, only two were from polygamous families, and that child abuse in Saudi Arabia is rare while polygamy is common. For those obsessed with scientific objectivity, it seems strange to me that such arguments, which could not pass by even the most lax science reviewer, did not stimulate their critical faculties. On the other hand, I think it will be interesting for Dr. Shelleh and colleagues to know that recently, we have been able to show that women of polygamous families in Syria were more likely to have mental distress or to be physically abused, than women of monogamous families, after adjusting for other sociodemographic factors such as age, age of marriage, education, economic status, etc.^{1,2}

In contrast to the statement by Shelleh et al. that polygamy was the main focus of my letter, the reader can readily notice that my main concern was the failure of medical and law enforcement personnel involved in the reported cases to intervene to protect the children and bring the perpetrators to justice, when they had multiple opportunities to do so. It was also a cry for the medical community to take an active role in cases of child abuse, and in the case of current interest, to try to investigate the destiny of the remaining victim to protect him if he is still alive. I am really disturbed by the fact that in the response of Dr. Shelleh et al., authored by six practicing medics and a senior judge, there was not a single word about any steps taken by them or others to investigate the fate of the remaining unfortunate child, or to bring those responsible for these sad events to justice. Science is an objective study of life, yet better science is an objective study of life in the face of unfavorable sociocultural circumstances.

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Hb Bart's Levels in Cord Blood in Hb H Disease: Follow-up of Cases with "Probable" Hb H Disease

To the Editor: Hb Bart's levels of 2%-8% in cord blood are seen in α -thalassemia minor, and it is widely accepted that Hb Bart's of >10% is diagnostic of Hb H disease.^{1,2}

TABLE 1. Red cell indices and Hb electrophoresis results in "probable" Hb H disease at birth and at 3 years of age.

No.	Hb g/dL	RBC x10 ¹² /L	MCV fl	MCH pg	RDW SD	Bart's %	H%	A%	F%	S%	A ₂ %
1	11.7	4.62	95.9	25.3	69.3	16.1	–	24	59.9	–	– (cord)
	10.0	5.87	56.9	17	37.5	–	–	96.7	–	–	3.3 (3 years old)
2	13.2	5.37	83.6	24.6	69.3	11.7	–	19.1	69.2	T	– (cord)
	8.3	4.92	52	16.9	33.7	–	–	74.8	–	21.6	3.6 (3 years old)
3	Clot	Clot	80	30	52.1	12	–	20.3	61.2	6.5	– (cord)
	10.6	6.07	55	17.5	33.5	–	–	73.7	–	24.2	2.1 (3 years old)
4	10.3	3.7	105.9	27.8	96.5	10.9	–	25.1	63.9	–	– (cord)
	6.4	2.86	77.6	22.4	–	1.8	T	82.2	15.8	–	T (3 years old)

T=traces.

However, it has been reported by Pembrey et al.³ that Hb Bart's of even up to 16% might be present in severe cases of α -thalassemia in Eastern Saudi Arabia. On the basis of the above reports, we had classified four newborns (Table 1) whose cord blood Hb Bart's ranged from 10% to 16.1% as "probable" Hb H disease. We now present their follow-up findings at about three years of age.

Discussion

In our study, only 1 out of the 4 cases of "probable" Hb H disease was shown to have Hb H disease (Table 1, no. 4). The patient developed pancytopenia and has been dependent on transfusion for the last 2½ years. The patient has been referred for bone marrow transplant. It is evident that the majority of the newborns with Hb Bart's of 10%-16.1% do not develop probable Hb H disease and are cases of severe α -thalassemia trait. Exceptionally high prevalence of the α -^{T Saudi} mutation has been reported from Bahrain, which is geographically and ethnically closely related to Eastern Saudi Arabia.⁵ It is likely that our cases of severe thalassemia trait resulting in high Hb Bart's have an underlying compound heterozygosity for α -^{T Saudi} mutation and some other deletional α -thalassemia. In the absence of

molecular studies, cases with Hb Bart's of 10%-20% in cord blood should not be labeled as Hb H disease, and such samples should be followed up by repeat electrophoresis at 4-6 months of age to ascertain the correct phenotype.

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