tumours represent only 25% of all cardiac tumours. The most prevalent are metastatic cardiac tumours and pseudotumors. Estimated frequency of primary tumours of the heart ranges from 0.0017% to 0.33%.[2] Seventy-five percent of primary cardiac tumours are benign. Myxomas account for nearly half of them. Primary malignant cardiac tumours are predominantly sarcomas.[2]

Since noninvasive diagnostic modalities have become more sensitive, there has been marked increase in the number of patients who receive diagnoses.

Presenting symptoms, treatment options and prognosis are largely controlled by the anatomic location of the tumour. Tumours of the heart are known to be great mimickers.[2,3] Most patients with malignant cardiac tumour die within one year of initial diagnosis, either because of the often asymptomatic presentation of cardiac tumours until advanced disease or because of a low index of suspicion on the part of the physician. The survival rates for extracardiac malignancies have improved substantially. Meanwhile, the prognosis in cardiac malignancies is still dismal. Median survival for patients with cardiac sarcomas is 6 months, and 93 months for those with noncardiac sarcomas. Patients with primary cardiac sarcomas who undergo surgical resection have a median survival of 12 months. Patients who undergo only palliative chemotherapy have a median survival of one month.[4]

Management options for cardiac tumours include surgery, neoadjuvant and adjuvant chemotherapy. Surgical resection is the treatment of choice.[5] In the case of inoperable disease (i.e., unresectable tumour, presence of metastases) palliative chemotherapy should be offered, although in some cases, palliative surgical debulking may be undertaken to relieve rapidly progressing symptoms. Younger patients with no metastatic disease may be considered suitable candidates for orthotopic heart transplant.[4]

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Oculopharyngeal muscular dystrophy: Oculopharyngeal muscular dystrophy (OPMD, OMIM 164300) fits the description of a neglected condition as described in the *CMAJ* editorial by Kelsall.[1]

This condition has escaped attention in many medical school and resident teaching curricula despite having an unusually high prevalence in those with Quebec French–Canadian ancestry. This is due to a founder effect. A mutant trinucleotide repeat expansion allele was introduced by three emigrant French sisters in 1648.[2] The most common disease manifestations are dysphagia, ptosis and limb weakness, with symptoms usually appearing in the fifth decade. The disease often goes undiagnosed when referral for dysphagia, which can be severe, is made. A label of myasthenia gravis is occasionally incorrectly applied as well. Treatment can be difficult, but is available for both dysphagia and ptosis. Counselling patients on the hereditary nature of this condition (autosomal dominant) frequently results in identification of additional affected family members.

Greater awareness would improve diagnosis and mitigate the unnecessary investigation that frequently occurs, including repeated upper endoscopy. A Scottish study showed a 3-to-20-year delay from symptom onset to diagnosis, with a quarter of patients with dysphagia having undergone a decade of investigation and treatment for pharyngeal problems.[3] The findings are congruent with the experience shared by many who manage these patients. Because patients of Quebec French–Canadian ancestry represent one of the world's most prevalent OPMD carrier groups, this disease should receive more attention in our country.

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Some letters have been abbreviated for print. See www.cmaj.ca for full versions and competing interests.

Correction

Author's affiliation in Mar. 4, 2014, issue

In the *CMAJ* article "The association between ownership of common household devices and obesity and diabetes in high, middle and low income countries,"[1] the affiliation for Roya Kelishadi should be "Isfahan Cardiovascular Research Center, Isfahan Cardiovascular Research Institute, Isfahan University of Medical Sciences, Isfahan, Iran.

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