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CASE REPORT

# The sick LADy who cried wolf: A case of Wellens' syndrome presenting in the shadow of chronic sickle cell pain

*La femme souffrante qui criait au loup: un cas de syndrome de Wellens dissimulé par des douleurs chroniques provoquées par la drépanocytose*

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**Abstract** **Introduction:** Chest pain is a common presenting complaint in the emergency centre (EC), with a very wide differential diagnosis. Evaluation of patients with chest pain in the EC is geared toward rapidly identifying and treating the subset of patients with potentially life-threatening causes, including acute coronary syndromes. ECG and cardiac biomarkers are indispensable tools in this endeavor.
**Case report:** A 47 year old African woman presented to the EC with chest pain; her ECG revealed findings typical of Wellens' syndrome. Subsequent coronary angiography revealed near-total proximal LAD occlusion.
**Discussion:** Wellens' syndrome refers to a pre-infarction stage of acute coronary syndrome with distinct ECG T-wave changes that strongly predict the presence of critical LAD coronary artery stenosis; it is a harbinger of impending extensive anterior myocardial infarction. Emergency physicians must be able to recognize its ECG features and institute appropriate intervention.

**Abstract** **Introduction:** Les douleurs poitrinaires sont une affection fréquemment enregistrée dans les services d'urgence, et sont associées à des diagnostiques différentiels très variés. L'évaluation des patients souffrant de douleurs poitrinaires dans les services d'urgences a pour objectif d'identifier et de traiter rapidement le sous-ensemble de patients souffrant de troubles potentiellement mortels, et notamment de syndromes coronariens aigus. Les électrocardiogrammes et biomarqueurs cardiaques sont des outils indispensables à ces efforts.
**Rapport d'enquête:** Une femme de 47 ans, d'origine africaine, s'est présentée au service d'urgences, souffrant de douleurs poitrinaires; son électrocardiogramme a permis de conclure à un syndrome de Wellens. Une angiographie coronaire subséquente a révélé une occlusion quasi-totale de l'artère antérieure gauche descendante.
**Discussion:** Le syndrome de Wellens est une étape pré-infarctus du syndrome coronarien aigu, associé à des changements distincts dans l'onde T de l'ECG, prédisant fortement la présence d'une sténose de l'artère coronaire antérieure gauche descendante; ceci révèle l'imminence d'un infarctus majeur du myocarde.

## African relevance

- Many parts of Sub-Saharan Africa are undergoing rapid urbanization, accompanied by widespread adoption of Westernized lifestyles.
- Practitioners in Sub-Saharan Africa will need to recognize the myriad of presentations of acute coronary syndromes.
- 12-lead ECG is an essential test in the risk stratification of patients complaining of chest pain and emergency physicians must be proficient in interpreting ECGs.

## Introduction

Chest pain is a frequent reason for emergency centre (EC) visits. Although it has an extensive differential diagnosis, algorithms for evaluating chest pain in the EC are generally geared toward ruling out potentially life-threatening causes, including acute coronary syndrome (ACS). In the work up of suspected ACS, clinical history, electrocardiogram (ECG), and cardiac enzymes are indispensable tools for risk stratification. Emergency physicians must be highly proficient in ECG interpretation to avoid missing potentially life-threatening diagnoses.

Wellens' syndrome (WS) refers to a pre-infarction variant of ACS with distinct ECG findings that if overlooked portends a grave outcome. It was originally described in 1982 as a distinct pattern of precordial T-wave abnormalities (biphasic T waves in anterior precordial leads V1-V3 or deeply inverted symmetric T waves in V1-V3), which are strongly predictive of critical left anterior descending (LAD) coronary artery stenosis.[1] Its significance lies in the fact that without urgent

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revascularization, these patients tend to progress to anterior myocardial infarction (MI) within a median time of about eight days, even with medical therapy.[1,2] Additionally, stress testing is contraindicated in these patients as this can precipitate fatal MI.[3] Once recognized, patients with WS must undergo urgent cardiac catheterization and revascularization. We present a case of WS in a black African female.

## Case report

A 47 year old black Zimbabwean female with a past medical history significant for sickle cell disease with frequent EC visits for "sickle cell pain" and hyperlipidaemia presented to the EC with 1 week of intermittent retrosternal chest pain. She described the pain as a dull, non-exertional pressure-like retrosternal discomfort; it did not radiate to the arms or neck and was not associated with dyspnoea or diaphoresis. The pain lasted about an hour and had resolved en route to the EC. Of note, the week prior to her current presentation, she had been seen three times in two different ECs with the same pain. She reported that three days ago she had an ECG done while having chest pain, which was normal. Her pain was attributed to her sickle cell disease and she was discharged with analgesia; this had not relieved her pain.

Family history was negative for premature coronary artery disease (CAD) or sudden cardiac death. She was a non-smoker and denied any recreational drug use.

Physical examination revealed an overweight female in no acute distress with stable vitals (BP 126/74 mmHg, pulse 86/min, RR 16/min, temperature 36.2 °C, SaO2 98% on room air). She had normal heart sounds, no murmurs, non-displaced cardiac apex, no JVD and no peripheral edema. The chest pain was not reproducible with palpation. Her breathing was non-labored with good diaphragmatic excursion, normal breath sounds and no wheezing or crackles. The remainder of her physical examination was unremarkable.

CBC and BMP were normal. Cardiac enzymes revealed a troponin I level $<0.10$ and CK-MB level of 0.76 ng/ml, both within normal limits. CXR showed normal cardio-mediastinal silhouette with normal lung fields. Her ECG is shown below in Fig. 1.

ECG findings were recognized as being typical of Wellens' syndrome. She was given 325 mg aspirin and 300 mg clopidogrel, started on a heparin infusion and transferred 300 km away for urgent coronary angiography.

Cardiac catheterization revealed 90–95% proximal LAD stenosis and 40% tubular eccentric lesion in the right circumflex artery; left circumflex artery was angiographically normal. Percutaneous transluminal angioplasty was performed with insertion of a drug-eluting stent. She was discharged uneventfully two days later on aspirin, clopidogrel, simvastatin, lisinopril and carvedilol.

## Discussion

As Sub-Saharan Africa is undergoing rapid urbanization, we are witnessing an accompanied profound adjustment in lifestyle with adoption of what has been traditionally referred to as a "Westernized lifestyle." This is characterized by reduced physical activity and diets rich in total calories, saturated fat, and refined food. Partly as a result of this, the epidemiology of disease in Sub-Saharan Africa is evolving with a rapidly rising burden of non-communicable diseases, including ischemic heart disease.[4] Health workers in Sub-Saharan Africa will increasingly be called upon to recognize and manage the different presentations of CAD. We report a case of Wellens' syndrome in a black Zimbabwean female.

In their seminal 1982 paper, *Wellens* et al. identified a subgroup of patients presenting with unstable angina (UA) with a specific ECG pattern that was strongly predictive of critical proximal LAD stenosis.[1] Out of 145 patients hospitalized for UA, they identified this ECG pattern, now widely known as

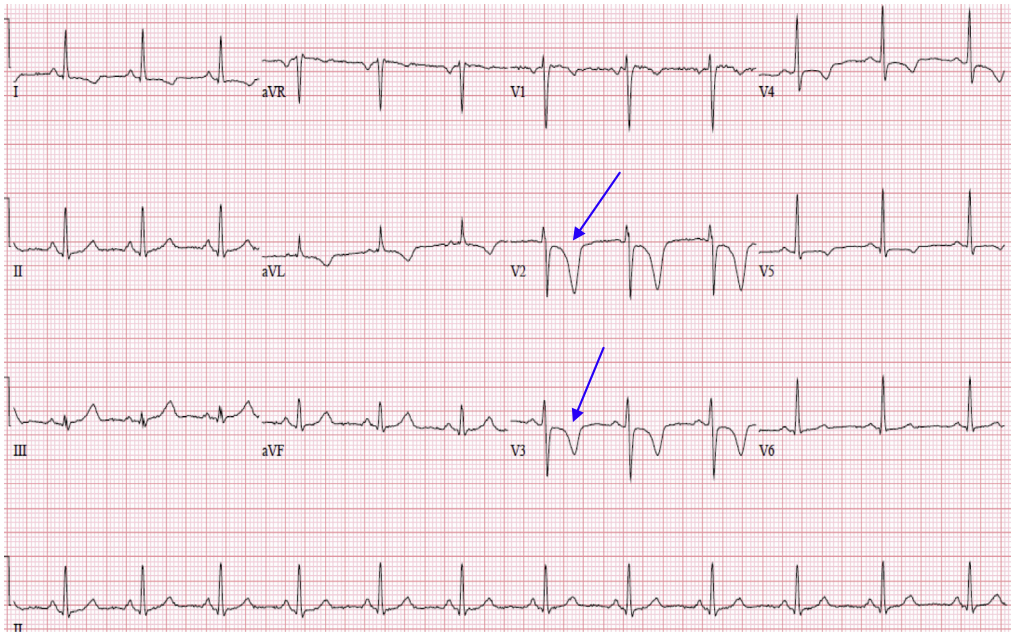


**Figure 1** ECG performed on presentation to EC, showing symmetrical deep T wave inversion in leads V1-V3 (typical of Wellens' syndrome type I).

Wellens' syndrome in 26 patients (18%). They also found that 12/16 WS patients (75%) who were managed medically without coronary revascularization progressed to extensive anterior MI within a mean time of 8.5 days.[1] A subsequent study of 1260 patients presenting with UA found that 180 (14%) had WS. Analysis of their angiographic findings revealed that they all had at least 50% stenosis of LAD, with an average of 85% stenosis.[2] Clearly, without revascularization, WS carries an extremely high risk of MI and sudden death. Given the poor prognosis of proximal LAD stenosis, it is known colloquially as the "widow maker lesion."

The diagnostic criteria of WS are shown in Table 1.

Two types of WS syndrome have been described: type I WS accounts for approximately 75% of cases and is defined by deeply inverted symmetric T waves in leads V1-V4, while type II WS accounts for approximately 25% of cases and is characterized by biphasic T-waves in the precordial leads, especially V2-V3, as illustrated in Fig. 2.

WS is truly a "wolf in sheep's clothing," a harbinger of death that can be easily missed for many reasons. Firstly, the ECG changes are subtle and easily overlooked by the unsuspecting eye. Many times the machine only reports this as "non-specific T wave changes," which are often ignored as benign findings. Added to this, when an ECG is obtained during periods of chest pain it can be totally normal. Classically, the ECG features of WS are manifest during a pain-free period; they can therefore be missed if one does not remember to repeat the ECG during a pain-free period. This is worth remembering, especially in settings where the instinct may be to conserve limited resources. To make matters worse, cardiac biomarkers are usually normal, which is often falsely reassuring. Published reports show that WS not uncommonly occurs even in young patients even in their twenties-thirties, often with no traditional cardiac risk factors, whom physicians would least expect to have CAD.[5–7] To complicate matters further, the usual recourse of recommending exercise stress testing for chest pain patients to rule out CAD may turn out to be a death sentence for a patient with WS; provocative testing is absolutely contraindicated as it has been known to precipitate anterior MI with a potentially fatal outcome.[3,8] To top it off, physicians do not have the luxury of time or a second chance to make the diagnosis as WS can progress to extensive anterior MI within a short period of time. There have been case reports of this happening within an hour while the patient was still in the EC[9]; in fact, the most rapid progression to MI reportedly happened within 7 min.[10] All these factors underline how crucial it is to recognize the ECG findings and institute timely invasive intervention.

As far as we know, this is the first report of WS in a black African patient. Interestingly, she had a normal ECG during a period of chest pain three days prior to her presentation. This may have been because of transient pseudo-normalization of

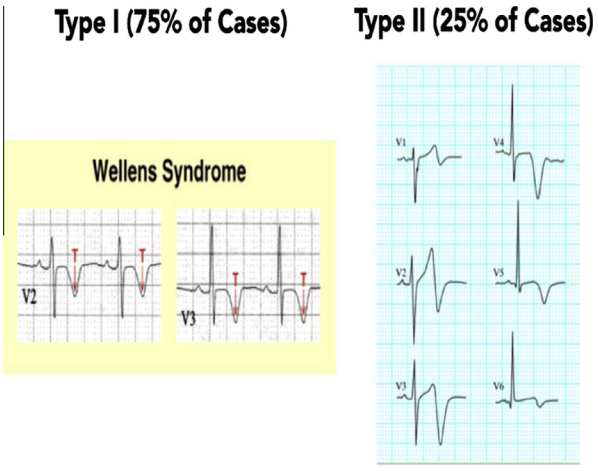


**Figure 2** Schematic showing typical EKG patterns of Wellen's syndrome.

Wellens' changes during chest pain. Had a serial ECG been done during a pain-free period, the T-wave changes would likely have been unmasked. Her long-standing history of sickle cell disease was also a confounding factor as her pain was attributed to sickle cell pain.

Among the many possible causes of T-wave inversions in anterior leads that can mimic WS, in a young African female "persistent juvenile pattern" can be challenging to differentiate from WS; anterior lead T-wave inversions are reportedly common in young black women and this represents a benign variant of normal.[11] The crucial difference is that while Wellens' T-wave inversions are deep and symmetric, the T wave changes seen with persistent juvenile pattern are shallow and asymmetric. In settings where cardiac catheterization is not readily available and patients have to be referred at a great cost over long distances to larger centers, this distinction is crucial. In our case, we were able to use SMS picture messaging to consult an experienced cardiologist in the US by sending the ECG for a second opinion. With wider access to cellphones and internet, such applications of Telemedicine may help practitioners in remote areas seeking consultation with experts who are often based in larger cities.

## Conclusion

The incidence of CAD is rapidly rising in Sub-Saharan Africa in part due to rapid urbanization and the attendant lifestyle changes. Practitioners in Sub-Saharan Africa will increasingly be called upon to recognize the various presentations of ACS. WS is a subtype of UA with characteristic ECG changes strongly associated with critical stenosis of the proximal

**Table 1** Wellens' syndrome criteria.

- Prior history of chest pain
- Normal ECG during chest pain or only mild ST elevation or depression, or with terminal negative deflection of the T wave in V1 and V2
- Normal or mildly elevated cardiac enzymes
- No pathologic precordial Q waves
- Normal precordial R wave progression
- Deeply inverted or biphasic T waves in V2 and V3, possibly V1, V4, V5 and/or V6 during a pain-free interval

LAD. Once recognized, patients require urgent revascularization to prevent potentially fatal anterior myocardial infarction.

## Conflict of interest

The authors declare no conflict of interest.

## References


1. de Zwaan C, Bar FW, Wellens HJ. Characteristic electrocardiographic pattern indicating a critical stenosis high in left anterior descending coronary artery in patients admitted because of impending myocardial infarction. *Am Heart J* 1982;**103**(4 Pt ):730–6.
2. de Zwaan C, Bar FW, Janssen JH, Cheriex EC, Dassen WR, Brugada P, et al. Angiographic and clinical characteristics of patients with unstable angina showing an ECG pattern indicating critical narrowing of the proximal LAD coronary artery. *Am Heart J* 1989;**117**(3):657–65.
3. Tandy TK, Bottomy DP, Lewis JG. Wellens' syndrome. *Ann Emerg Med* 1999;**33**:347–51.
4. Noncommunicable diseases. 2013 World Health Organization report online cited 2013 August 7. Available from: http://www.who.int/mediacentre/factsheets/fs355/en/(2013).
5. Mead NE, O'Keefe KP. Wellen's syndrome: an ominous EKG pattern. *J Emerg Trauma Shock* 2009;**2**:206–8.
6. Khan B, Alexander J, Rathod KS, Farooqi F. Wellens' syndrome in a 24-year-old woman. *BMJ Case Rep* 2013;**2013**.
7. Lilaonitkul M, Robinson K, Roberts M. Wellens' syndrome: significance of ECG pattern recognition in the emergency department. *Emerg Med J* 2009;**26**:750–1.
8. Rhinehardt J, Brady WJ, Perron AD, Mattu A. Electrocardiographic manifestations of Wellens' syndrome. *Am J Emerg Med* 2002;**20**:638–43.
9. Donahue B, Chan SB, Bhandarkar S. Rapid progression of Wellens syndrome in the emergency department. *J Emerg Med* 2012;**43**:667–70.
10. Stankovic I, Vlahovic-Stipac A, Ilic I, Putnikovic B, Neskovic AN. Aborted myocardial infarction in a patient with rapid progression of Wellens syndrome. *J Emerg Med* 2012;**43**:e181–4.
11. Roukoz H, Wang K. ST elevation and inverted T wave as another normal variant mimicking acute myocardial infarction: the prevalence, age, gender, and racial distribution. *Ann Noninvasive Electrocardiol* 2011;**16**:64–9.