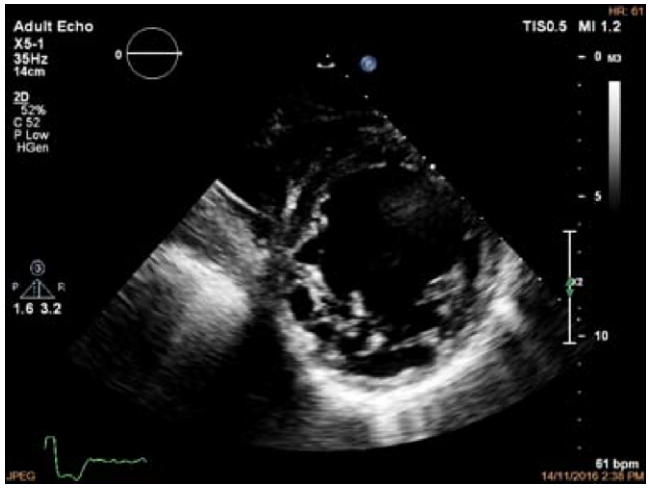

At 33 years of age he underwent successful valve repair for severe mitral regurgitation.

Follow-up echocardiography showed severe left ventricular dilatation and severe systolic dysfunction with hyper-trabeculation of the lateral wall, suggestive of isolated ventricular non–compaction. Anti–heart failure treatment was optimised and atypical causes of heart failure were considered. Genetic testing was performed on a panel of 174 cardiac disease genes. This identified a heterozygous variant in the *COL3A1* gene (c.2959G>A, p.Gly987Ser), confirming a diagnosis of vascular Ehlers-Danlos syndrome.

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0060

A Curious Case of Environmental Cardiomyopathy

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We present a unique case of a temporal relationship of cardiomyopathy in an unrelated husband and wife couple. A 39-year-old man presented to our institution with a subacute history of dyspnoea, with clinical features of biventricular heart failure. There were no antecedent triggers bar a remote history of a viral infection several months prior and the patient had no family history of cardiac disease. Transthoracic echocardiography demonstrated severe biventricular systolic impairment (left ventricular ejection fraction 17%) with evidence of severe spontaneous echo contrast in the left ventricle. Coronary angiography revealed minor coronary disease. Right ventricular biopsy revealed myocyte hypertrophy with interstitial fibrosis. He was treated with heart failure-specific therapy and discharged for follow-up. Unfortunately, his 28-year-old wife, who was >6 months postpartum, had a similar presentation to our institution with examination findings of right-sided heart failure 2 months after the husband's index presentation. Similarly, the only antecedent triggers identified were of a possible viral illness several months predating her presentation. Transthoracic echocardiography revealed severe biventricular systolic impairment (left ventricular ejection fraction 23%). Coronary angiography revealed normal coronary arteries. Right ventricular biopsy similarly revealed mild myocyte hypertrophy with mild interstitial fibrosis. She was treated with heart failure-specific therapy and similarly discharged. Both of them were given a diagnosis of idiopathic dilated cardiomyopathy. This case emphasises the importance of potential environmental factors in the aetiology of idiopathic cardiomyopathy and the value of further research to establish incident triggers and underlying disease substrates.

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0061

A Randomised Pilot Study of Chewing Gum to Relieve Thirst in Chronic Heart Failure (RELIEVE-CHF Trial)

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Background: Thirst is a burdensome symptom of chronic heart failure (CHF). To date, there remains limited scientific evidence to advise clinicians on how to manage thirst in daily clinical practice. RELIEVE-CHF is the first trial to investigate the use of chewing gum to relieve thirst in CHF. The aim of this study was to determine the effect of chewing gum on the level of thirst, weight stability, and quality of life on individuals with CHF.

Methods: A total of 71 individuals with CHF on oral loop diuretics were recruited from St. Vincent's Hospital Sydney. Thirty-six participants were randomised to chewing gum (54 ± 14 years; 61% men) and 35 to no chewing gum (54 ± 15 years; 66% men) for 2 weeks. Both groups were assessed for their level of thirst (using a visual analogue scale and numeric rating scale) and weight at days 1–4, 7, 14, and 28, and quality of life at day 28.

Results: Those who received chewing gum had a statistically significant improvement in the level of thirst at day 4 (Figure 1A [$p = 0.04$]; Figure 1B [$p = 0.019$]) and day 14 (Figure 1C [$p = 0.02$]; Figure 1D [$p = 0.021$]) compared to those who did not. No group differences were observed in weight over the study period and quality of life at day 28.

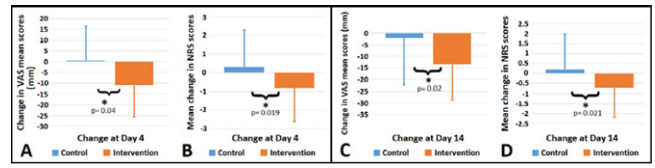

Figure 1.

Conclusion: Chewing gum provided relief from thirst but did not influence weight or quality of life.

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0062

A Rare Case of Infiltrative Cardiomyopathy Secondary to Scleromyxoedema

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A 44-year-old woman with a history of treatment-refractory scleromyxoedema presented with clinical heart failure along with dyspnoea and orthopnoea. Her previous transthoracic echo (TTE) 3 months prior showed normal left ventricular (LV) and right ventricular (RV) function with a stable pleural effusion. She had a raised N-terminal pro-B-type natriuretic peptide and creatinine kinase of 773 pmol/L and 4534 U/L, respectively. Her TTE showed new biventricular dysfunction, with diffuse hypokinesis and a LV ejection fraction (EF) of 20–25%. She underwent a coronary angiogram with right heart studies, which ruled out ischaemic cardiomyopathy or pulmonary hypertension. A cardiac magnetic resonance imaging (cMRI) confirmed biventricular failure (LVEF 37%; RVEF 28%) with the known stable pleural effusion (Figure 1A). Late gadolinium-enhanced images on cMRI showed diffuse patchy enhancement consistent with infiltrative disease. An endomyocardial biopsy showed myocarditis with interstitial fibrosis. There was polymorphous inflammatory infiltrate with accompanying myocyte damage. Interstitial mucin deposition was later confirmed with a colloidal iron stain, a hallmark of scleromyxedema. A thigh MRI and biopsy showed a necrotising myositis, confirming systemic disease. She responded to high-dose prednisolone, thalidomide, and intravenous immunoglobulin. Her follow up TTE showed stable LV function and improved RV function.

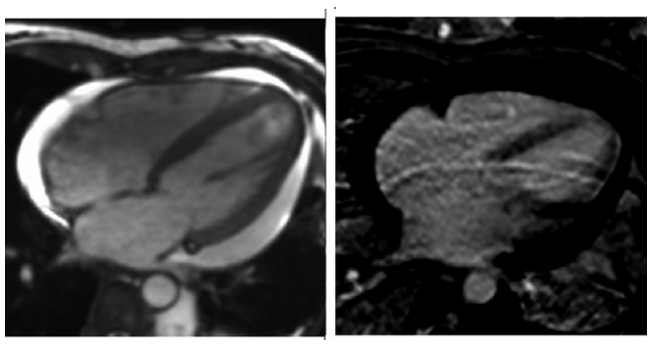

Figure 1.

This is the first documented case of biventricular failure secondary to scleromyxedema, with correlating endomyocardial biopsy and cMRI results. This case illustrates the diagnosis and possible treatment of a rare infiltrative cardiomyopathy, for which evidence in current literature is scarce.

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0063

This abstract has been withdrawn.

0064

A Rare Presentation of Reverse Takotsubo Cardiomyopathy with Acute Porphyria

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A 32-year-old woman had multiple presentations to the emergency department with severe abdominal pain and vomiting in context of a previous diagnosis of endometriosis with multiple endometrial ablations. Cardiology was consulted for sinus tachycardia. She had no electrocardiogram changes and had an initial Hs-trop of <5, peaking later at 200. An echocardiogram revealed severe left ventricular dysfunction with apical hyperkinesis and circumferential akinesis of the mid and basal ventricle. Cardiac magnetic resonance imaging revealed no evidence of myocardial oedema or fibrosis. Computed coronary angiogram was entirely normal. In the interim, the patient had an extensive work-up and multidisciplinary reviews for her abdominal pain, with no obvious cause. The diagnosis was suspected when a urine sample awaiting collection by the bedside turned red. Urinary porphobilinogen and porphyrins were highly elevated. Molecular genetic testing confirmed *HMBS* mutation, associated with acute intermittent porphyria. The patient was started on intravenous haema-arginate with resolution of symptoms. Serial transthoracic echocardiograms over the next 2 weeks revealed normalisation of left ventricular function. The patient continues to receive human hemin injections regularly and has had no further cardiac issues.

Conclusion: Reverse Takotsubo is a rare variant of Takotsubo cardiomyopathy, forming only 2.2% of the cases reported in the International Takotsubo Registry [1]. This case highlights an atypical presentation of a rare condition in context of an exceedingly rare condition. Methodical work-up and attention to detail have yielded an important diagnosis after remaining elusive for many years.

Reference

[1] Templin C, Ghadri JR, Diekmann J, Napp LC, Bataiosu DR, Jaguszewski M, et al. Clinical features and outcomes of Takotsubo (stress) cardiomyopathy. N Engl J Med 2015;373:929–38.

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