Results

Total of 81 patients were recruited. Fifty-four patients were on ARNI and 27 on ACEi/ARB. Baseline characteristics such as co-morbidities and medications used were similar between two groups. There was a statistically significant improvement of ejection fraction in ARNI group: pre-treatment (EF 32.4%) vs follow up (EF 36.85%; $p < 0.001$). LVIDd, LVIDs, and LVEDV also show statistically significant changes in the ARNI arm (5.76 cm vs 5.60 cm [$p = 0.0042$], 4.84 cm vs 4.46 cm [p = 0.004], and 103.85 ml vs 93.59 ml [$p = 0.016$]). All study patients had significant improvement in NYHA class after treatment: ARNI $p < 0.001$ (28 patients); non-ARNI $p = 0.031$ (6 patients). However, for the median difference in length of admission within the first 90 days between treatment, there is a trend following the ARNI over the non-ARNI group 1.32 vs 10.06 with $p = 0.05$. Patients who received ARNI had statistically significant lower value of NT pro-BNP (1459 vs 3148; $p = 0.012$) and better quality of life (KCCQ: 82.81 vs 54.17; $p < 0.001$).

Conclusion

This showed improvement in EF, LVIDd, LVIDs, LVEDV after at least 3 months of ARNI. This study's results suggest that ARNI treatment had a better reverse cardiac remodeling effect compared to non-ARNI treatment. Patients who received ARNI also had a better quality of life (KCCQ) and a lower value of NT pro-BNP.

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Prevalence of acid alpha-glucosidase (GAA) pseudodefiency allele and it's clinical significance among patients with cardiomyopathy

H.B. CHOW[A], H.S. LING[A], E.Z. CHAN[A], B.K. CHUNG[B], L.K. THEN[B], I.X. PANG[B], H.K. KHOR[B], S.T. CHAN[A], C.T. TAN[B], K.T. KOH[B], Y.L. CHAM[B], A. SAID[A], A.Y.Y. FONG[B,C], T.K. ONG[B], Y.Y. OON[B]

[a]*Faculty of Medicine and Health Sciences,University Malaysia Sarawak, Sarawak, Malaysia*
[b]*Department of Cardiology, Sarawak Heart Centre, Sarawak, Malaysia*
[c]*Clinical Research Centre, Sarawak General Hospital*

Background

Pompe disease is an autosomal recessive lysosomal storage disorder caused by deficiency of lysosomal acid alpha-glucosidase (GAA) activity, leading to the progressive accumulation of glycogen in lysosomes of the skeletal and cardiac muscles. An alpha-glucosidase (GAA) pseudodeficiency allele is a change in the GAA gene sequence that results in GAA enzyme activity reduction, but does not cause Pompe disease. In Japan and Taiwan, there is high prevalence of pseudodeficiency allele (c.1725G > A and c.2065G > A) detected from their newborn screening. We observed similar prevalence of pseudodeficiency allele among our patients who had genetic test performed for suspected hereditary cardiomyopathy.

Objectives

To report the prevalence of GAA pseudodeficiency allele, and to ascertain its clinical significance among patients with cardiomyopathy.

Methods

The clinical data of the patients with GAA mutations were retrieved. Patients were called back for neurological examination, lung function test, measurement of creatine kinase (CK) level and dried-blood-spot for GAA enzymatic activity.

Results

From January to December 2021, 33 patients underwent genetic testing. 23 out of the 33 genetic analyses included GAA mutation. 9 (39.13%) out of 23 were tested positive for pseudodeficiency allele. Their median age was 53 years (range 29–82), 44.4% were males with equal ethnic distribution (33.3% Malay, 33.3% Chinese, 33.3% Dayaks). All were heterozygous for the pseudodeficiency allele: 5 (55.6%) with c.[1726A; 2065A] allele, the other 4 (44.4%) c.2065G > A. The underlying cardiomyopathy phenotypes were hypertrophic (44.4%), transthyretin amyloid (22.2%), hypertensive (22.2%) and Fabry (11.1%). 1 patient (11.1%) with transthyretin amyloid cardiomyopathy died of advanced heart failure at age 79 years. 1 patient had mild motor weakness of the limbs attributable to thyrotoxicosis, while the other 7 patients had normal skeletal motor function. Their median predicted forced vital capacity was 87.5% (range 76–103), median CK level was 103 U/L (range 39–297) and median GAA activity was 4.8 mmol/l/h (range 3.2–9.2) [normal >2.0].

Conclusion

The prevalence of GAA pseudodeficiency allele among patients with cardiomyopathy is 39.13%. None of the patients exhibit significant muscle weakness or respiratory insufficiency despite low normal enzymatic activity. Whether the presence of pseudodeficiency allele affects the prognosis of the underlying cardiomyopathy remains uncertain.

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Levosimendan versus dobutamine in acute heart failure: Insight into short and long term mortality

W.J. QUAH, M. AKMA, T. DEVI, J. JAAFAR, H.B. KOH, C.K. TEOH, A. MOHD GHAZI

Cardiology Department, Institut Jantung Negara

Background

Acute decompensated heart failure (ADHF) leads to substantial mortality. Levosimendan, a calcium sensitizer improves myocardial contractility without increase in myocardial oxygen demand. However, there is lack of local data to support usage.

Objective

To assess efficacy and safety of intravenous infusion of levosimendan versus dobutamine in acute setting.

Materials & methods

All patients in our ADHF registry were screened. Propensity score matching was performed to compare both arms of levosimendan and dobutamine. Multivariate logistics regression method and Kaplan Meier curves were constructed. Follow-up was until 52 weeks.

Results

156 patients were studied. 78 patients in the levosimendan arm versus 78 patients in the dobutamine arm. 42 patients in the levosimendan arm were also added on with dobutamine. 86% (67 patients) were male with mean age of 59.5 ± 12.7. 70% (55 patients) had CAD, 69% (54 patients) had DM and 40% (31 patients) had renal impairment. 46% of patients had systolic BP of less than 100 mmHg during initiation. Mean NT-proBNP levels were 12,079 pg/mL. More than 50% were on at least 2 of the GDMT. There was significant difference in regards to length of stay (LOS) between both