Factors influencing the choice-of-care pathway and survival in the fetus with hypoplastic left heart syndrome in New Zealand: a population-based cohort study

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ABSTRACT

Objectives To better understand the relative influence of fetal and maternal factors in determining the choice-of-care pathway (CCP) and outcome in the fetus with hypoplastic left heart syndrome (HLHS).

Design A retrospective, population-based study of fetuses with HLHS from a national dataset with near-complete case ascertainment from 20 weeks' gestation. Fetal cardiac and non-cardiac factors were recorded from the patient record and maternal factors from the national maternity dataset. The primary endpoint was a prenatal decision for active treatment after birth (intention-to-treat). Factors associated with a delayed diagnosis (>24 weeks' gestation) were also reviewed. Secondary endpoints included proceeding to surgical treatment, and 30-day postoperative mortality in liveborns with an intention-to-treat.

Setting New Zealand population-wide.

Participants Fetuses with a prenatal diagnosis of HLHS between 2006 and 2015.

Results Of 105 fetuses, the CCP was intention-to-treat in 43 (41%), and pregnancy termination or comfort care in 62 (59%). Factors associated with intention-to-treat by multivariable analysis included a delay in diagnosis (OR: 7.8, 95% CI: 3.0 to 20.6, p<0.001) and domicile in the maternal fetal medicine (MFM) region with the most widely dispersed population (OR: 5.3, 95% CI: 1.4 to 20.3, p=0.02). Delay in diagnosis was associated with Māori maternal ethnicity compared with European (OR: 12.9, 95% CI: 3.1 to 54, p=0.001) and greater distance from the MFM centre (OR: 3.1, 95% CI: 1.2 to 8.2, p=0.02). In those with a prenatal intention-to-treat, a decision not to proceed to surgery was associated with maternal ethnicity other than European (p=0.005) and the presence of major non-cardiac anomalies (p=0.01). Thirty-day postoperative mortality occurred in 5/32 (16%) and was more frequent when there were major non-cardiac anomalies (p=0.02).

Conclusions Factors associated with the prenatal CCP relate to healthcare access. Anatomic characteristics impact treatment decisions after birth and early postoperative mortality. The association of ethnicity with delayed prenatal diagnosis and postnatal decision-making suggests systemic inequity and requires further investigation.

INTRODUCTION

A diagnosis of hypoplastic left heart syndrome (HLHS) carries a high risk of mortality and morbidity. Despite steady improvements in perioperative care and operative techniques, it continues to be the most common cardiac anomaly associated with death in the first year of life. In New Zealand, the incidence of HLHS is 0.22/1000 births—similar to that in northern Europe when maternal ethnicity is European, and lower when maternal ethnicity is Māori or Pacific. Mortality from the first stage procedure to the second stage procedure was 17% between 1992 and 2014, similar to that reported elsewhere.

Antenatal detection of HLHS provides an opportunity for prenatal counselling. Issues discussed at that time include the risks and benefits of treatment and the impact of the treatment pathway on the mother and family. At that point, many will be in a position to...
make a choice on whether to continue with the pregnancy or have a termination of pregnancy, and whether to pursue surgical treatment after birth. Where active treatment is considered, birth can be arranged in a controlled environment at or near a surgical centre.

Though operative mortality is influenced by fetal and infant characteristics, factors impacting the choice-of-care pathway (CCP) following a prenatal diagnosis are poorly defined. In particular it is unclear whether a care pathway is chosen because of anatomic risk factors or for other reasons. In New Zealand, the diagnosis of HLHS in fetuses of Māori and Pacific mothers occurs later during pregnancy and these ethnic groups have lower termination rates and higher overall mortality. The cause of this inequity is unclear but could relate to access to healthcare, parental and family values, implicit bias or, given the heterogeneity of HLHS, differences in disease severity. Several recent reports have highlighted the impact of access on decision-making in this condition. A better understanding of this process has the potential to impact the future design of maternal, fetal and paediatric cardiac programmes in areas where there is inequity.

METHODS
Study design and case identification
New Zealand has a single fetal and paediatric cardiac centre based at Starship Children’s Hospital in Auckland. In addition, there are three maternal fetal medicine (MFM) hubs, Hub 1 at the cardiac centre, and Hubs 2 and 3 elsewhere in the country. At Hub 1, women with a fetus suspected of having HLHS are seen by an MFM specialist and a fetal cardiologist. In Hubs 2 and 3 they are seen by an MFM specialist with images subsequently forwarded to the fetal cardiologist for review (by DVD in the first 2 years of the study and by secure internet file transfer subsequently). Counselling is undertaken by video link with a fetal cardiologist and MFM specialist in attendance. Discussions include the possibility of active treatment, comfort care or termination of pregnancy. If the diagnosis is uncertain the woman is seen at the cardiac centre.

Study participants were identified from the national fetal cardiology database, the national cardiac surgical database, and from the Perinatal and Maternal Mortality Review Committee (PMMRC) dataset of perinatal deaths. It is a statutory requirement in New Zealand to report all perinatal deaths from 20 weeks’ gestation, including late terminations of pregnancy, to the PMMRC. Thus, the dataset has near-complete case ascertainment at a national level from that stage in pregnancy, with only the potential for un-reported cases missing. Demographic data were obtained from the Ministry of Health’s National Maternity Collection. Data were merged into a single dataset using mother and/or baby New Zealand National Health Index number as a unique identifier. Hospital records were reviewed to supplement information as needed.

All cases with a prenatal diagnosis of HLHS in New Zealand between January 2006 and December 2015 were included. Cases with an unbalanced atrioventricular canal defect or double outlet right ventricle were excluded. Those with extracardiac and chromosomal anomalies were included. The diagnosis of HLHS was considered to have occurred at the time it was confirmed at an MFM hub.

This population-based audit was approved by the Health and Disability Ethics Committees of New Zealand (MEC12EXP077AM06).

Ethnicity
In New Zealand, the collection of ethnicity data for health purposes involves individuals nominating all ethnic groups with which they identify. Where an individual identifies with more than one ethnic group, a single ethnicity is assigned using the prioritisation process recommended in the Ministry of Health’s ethnicity data protocol. Prioritised ethnic groups are categorised into five subgroups: Māori, Pacific Peoples, Asian, Middle Eastern, Latin American and African (MELAA), and European.

Socioeconomic status
The New Zealand Index of Deprivation (NZDep) is an area-based measure of socioeconomic deprivation that is calculated after each Census. Census meshblocks, a small geographical area with a population of around 60–110 people, are assigned a deprivation score derived from Census variables that include measures of income, welfare dependency, education, housing and access to transportation and communication. Meshblocks are then categorised into deciles according to their deprivation scores with about 10% of the New Zealand population residing within each decile. Decile 1 represents areas with the least-deprived households and 10 represents areas with the most-deprived households. Deciles were grouped into quintiles for analysis. NZDep 2006 was used for mothers delivering between 2006 and 2012, and NZDep for 2013 for those between 2013 and 2015.

Indices of disease severity
Cardiac anatomical factors were identified on the most recent fetal echocardiogram. Known cardiac anatomical factors associated with disease severity included: the combination of mitral stenosis and aortic atresia; a smaller ascending aorta diameter; a restrictive or intact atrial septum; tricuspid regurgitation and right ventricle (RV) dysfunction.

The ascending aorta diameter was expressed as a Z score relative to gestational age and was analysed both as a continuous and categorical variable with the cut-off set at −4. This value was chosen as it is equivalent to an
ascending aorta diameter of <4mm in the newborn. We have previously found this diameter to be associated with reduced survival after Stage 1 surgery.\textsuperscript{5} The diagnosis of an intact or restrictive atrial septum was based on fetal echocardiographic findings of a small or absent atrial communication and/or abnormal pulmonary venous Doppler (Type B or C by the criteria of Takezawa et al\textsuperscript{[4]}). The degree of tricuspid regurgitation was assessed qualitatively as absent, mild, moderate or severe and was grouped for analysis as none or mild versus moderate or severe. Likewise, RV dysfunction was assessed qualitatively and grouped as absent or present.

**Primary and secondary end points**

The primary endpoint was a decision to pursue active treatment following a prenatally diagnosed ascertained by review of the case records. Secondary endpoints were proceeding to cardiac surgery in liveborns with a prenatal intention-to-treat, and 30-day postoperative mortality in those who underwent cardiac surgery. A separate analysis was undertaken to determine factors associated with the gestational age at diagnosis, defined as first assessment by a MFM specialist and/or fetal cardiologist.

**Statistical analysis**

Frequency data are expressed as number and per cent, and compared across groups using $\chi^2$ and Fisher’s Exact tests (where expected cell count $<5$). Continuous data are expressed as median and range if non-parametric, or mean and SD if normally distributed, and compared using Wilcoxon rank-sum and Student’s t-test, respectively. Gestational age at diagnosis was dichotomised at 24 weeks as an a priori cut-off, reflecting viability and a point where termination of pregnancy was less likely to be an acceptable to women and clinicians. There were small numbers of missing data for cardiac anatomical measures. These are described in the tables, and are included with absence of the factor in statistical tests except for the continuous measure for ascending aortic diameter, where the individuals with missing data are excluded from the analysis.

Multivariable analysis was undertaken using logistic regression. The model exploring CCP included the following variables for a priori reasons: ethnicity, age, deprivation quintile (quintile 5 vs 1–4), gestation at diagnosis (<24 vs ≥24 weeks), cardiac risk factor indices, distance from the surgical centre (km), MFM hub and distance from the MFM hub (km). Variables were removed from the full model if they were not statistically significant and were not apparent confounders (ie, their removal from the model did not change the coefficients in the final model by >10%) by a process of manual step-wise removal. No interaction terms were defined.

The model predicting delay to diagnosis included all variables significantly associated with the outcome in the univariable analysis. Statistical significance was assumed at p<0.05.

**RESULTS**

**Cases**

Between January 2006 and December 2015, 117 fetuses and infants were identified with HLHS in New Zealand. Of these, 105 (90%) were diagnosed prenatally and form the basis of this report (figure 1). The remaining 12 were diagnosed after birth and included 8 live births, 2 of whom were diagnosed at postmortem, and 4 stillbirths.

In the 105 diagnosed prenatally, the median maternal age was 30 years and the median gestational age at diagnosis was 23 weeks (range 12–39 weeks) with 60 (57%) diagnosed before 24 weeks (table 1). Mothers identified as Māori in 19%, Pacific peoples 8%, Asian in 11% and European in 62%. None identified as MELAA. Thirty-one per cent were domiciled in the most deprived quintile, 59% lived more than 50 km from the surgical centre, and 36% lived more than 50 km from a MFM centre. Major non-cardiac anomalies were present in 18, including six with chromosomal anomalies. At least one cardiac risk factor was present in 69%.

**Antenatal CCP**

Termination of pregnancy occurred in 52 (50%). In an additional 10 (9%) cases there was a parental choice for comfort care. A greater proportion of those who had termination of pregnancy were diagnosed <24 weeks’ gestation (45/60 (75%) <24 weeks’ vs 7/45 (16%) ≥24 weeks’ gestation, p<0.001), while those with comfort care were less likely to be diagnosed <24 weeks gestation (2/60 (3%) < 24 weeks’ vs 8/45 (18%) ≥24 weeks, p=0.02). An active decision to pursue surgical treatment (intention-to-treat) was made in the remaining 43 (41%). Factors associated with this decision by univariable analysis included a gestational age at diagnosis of ≥24 weeks, younger maternal age, Māori and Pacific Peoples, the most deprived quintile, greater distance from the surgical centre (as a continuous variable), the MFM Hub (3 vs 1 and 2) and a larger ascending aorta Z score (table 1). RV dysfunction was more common in those cases where a decision was made to pursue surgical treatment. In a multivariable model, the decision to pursue surgical treatment was strongly predicted by later gestational age at diagnosis (OR: 7.8, 95% CI: 3.0 to 20.6, p<0.001) and MFM hub (Hub 3 vs 1 and 2, OR: 5.3, 95% CI: 1.24

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**Patient and public involvement**

No patients were involved.
Table 1  Factors associated with choice-of-care pathway among fetuses diagnosed with HLHS

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<th></th>
<th>Total</th>
<th>Termination or comfort care</th>
<th>Intention-to-treat</th>
<th>Univariable (p value)</th>
<th>Multivariable OR (95% CI)</th>
<th>P value</th>
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<td>62 (59)</td>
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<td>Gestational age at diagnosis (weeks) (median (range))</td>
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<td>22 (12–37)</td>
<td>29 (20–39)</td>
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<td>13 (22)</td>
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<td>≥24 weeks</td>
<td>45 (43)</td>
<td>15 (33)</td>
<td>30 (67)</td>
<td>&lt;0.001</td>
<td>7.8 (3.0 to 20.6)</td>
<td>&lt;0.001</td>
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<td>31 (6)</td>
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<td>0.9 (0.9 to 1.0)</td>
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<td>5 (36)</td>
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<td>16 (15)</td>
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<td>5 (31)</td>
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<td>16 (64)</td>
<td>9 (36)</td>
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<td>32 (36)</td>
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<td>3</td>
<td>17 (16)</td>
<td>6 (35)</td>
<td>11 (65)</td>
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<td>5.3 (1.4 to 20.3)</td>
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<td>20 (0–583)</td>
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<td>&gt;50 km</td>
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<td>19 (50)</td>
<td>19 (50)</td>
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<td>9 (50)</td>
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<td>−4.6 (3.1)</td>
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<td>2 (14)</td>
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<td>13 (87)</td>
<td>2 (13)</td>
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Values are n (%) unless otherwise stated. % are by column for total and by row for termination or comfort care and Intention-to-treat. P-values ≤ 0.05 are in bold.
HLHS, hypoplastic left heart syndrome; MFM, maternal fetal medicine; RV, right ventricle; TR, tricuspid regurgitation.
to 20.3, p=0.02). Lowest deprivation quintile tended to predict a decision to pursue surgical treatment (quintile 5 vs 1–4, OR: 2.7, 95% CI: 0.9 to 7.6, p=0.07). Neither the presence of major non-cardiac anomalies, nor cardiac anatomic risk factors were predictive of choice of care in the multivariable model.

**Timing of diagnosis**

Given the strong association between the CCP and gestational age at diagnosis, we reviewed factors that might be associated with a delayed diagnosis (diagnosis ≥24 weeks’ gestation) (table 2). A delayed diagnosis was strongly associated with maternal ethnicity, occurring in 17/20 (85%) of Māori, 4/8 (50%) of Pacific peoples, 4/12 (33%) of Asian and 20/65 (31%) of European pregnancies (p<0.001). It was more common in those who lived >50 km from an MFM centre (22/38, 58% vs 23/67, 34%, p=0.01) (figure 2). In a multivariable model, Māori maternal ethnicity (compared with European, OR: 12.9, 95% CI: 3.1 to 54, p<0.001) and domicile >50 km distance from the MFM centre (OR: 3.1, 95% CI: 11.2 to 8.2,
p=0.02) were independently associated with a delayed diagnosis.

**Live births and surgery**

Of the 43 fetuses with a prenatal intention-to-treat there were 37 live births and six stillbirths. Cardiac surgery was undertaken in 32 of the 37 live births, and consisted of a Norwood procedure in all 32 infants. All five of those who did not undergo surgery had additional cardiac or non-cardiac comorbidities that had not been identified, or the severity of which was not apparent, prior to birth. Factors associated with a decision to proceed to cardiac surgery in these 37 newborns included an absence of major non-cardiac anomalies, and ethnicity with surgical treatment occurring more frequently in infants whose mothers identified as European compared with Māori, Pacific peoples and Asian (p=0.005) (table 3). Multivariable analysis was not undertaken given the small sample size. Of the 32 who underwent cardiac surgery, 27 (84%) survived 30 postoperative days. Thirty-day mortality was more common in those with major non-cardiac anomalies (2/2 (100%) with major non-cardiac anomalies vs 3/30 (10%) without p=0.02), and was weakly associated with tricuspid regurgitation and RV dysfunction (both p=0.06).

**DISCUSSION**

In this population-based cohort of pregnancies and fetuses with HLHS and near-complete case ascertainment, the CCP was strongly associated with the gestational age at diagnosis. In a multivariable model, CCP was also associated with MFM region but not with indices of disease severity. A delayed diagnosis (≥24 weeks) was associated with Māori maternal ethnicity and distance from the MFM hub. After birth, the progression through the care pathway was modified by the presence of non-cardiac anomalies, and was also associated with maternal ethnicity.

The relationship between termination of pregnancy and gestational age at diagnosis is well described, particularly in jurisdictions where the availability of termination is limited by gestational age. In New Zealand, termination of pregnancy can be offered throughout pregnancy when there is a major congenital anomaly. Nevertheless, termination involves a feticide procedure from 22 weeks’
gestation. Moreover, termination at a later gestational age may be less acceptable to families and clinicians, and requires transfer to a supra-regional centre. Comfort care was the CCP in a minority of cases, and was chosen more frequently in those with a delayed diagnosis.

The CCP was also associated with an MFM hub remote to the surgical centre. While this association may reflect cultural or counselling differences, counselling was provided through a centralised videoconferencing facility by a fetal cardiologist and an MFM specialist. Of note, of the three MFM hubs this hub serves the most dispersed population (6.9 people/km² vs 16/km² and 39/km²) over the largest area (130000 km², vs 72000 km² and 62000 km²) and is situated furthest from the surgical centre. As such the association between MFM hub and the CCP may be a marker for rurality, identified by others to impair access to healthcare.

Anatomic factors were not important determinants of the CCP. A lack of association between disease severity and the decision to terminate a pregnancy has been reported previously, with similar rates of termination in fetuses with standard and higher-risk features.21,22

As a delay in diagnosis was strongly associated with the CCP, we reviewed factors that might be associated with this outcome. A delay in diagnosis was more common in pregnancies of Māori woman and in those domiciled >50 km from the MFM hub. There are a number of reasons why a diagnosis of HLHS may be delayed including: (1) delayed booking for maternity care, and/or delayed referral for the mid-trimester ultrasound screening examination; (2) a missed diagnosis at the screening ultrasound, and (3) a delay in referral or booking for a MFM consultation.

It is well documented that Māori experience inequities across almost all indicators of health status and in access to health services compared with Europeans.20,21 Māori and Pacific women have their initial obstetric assessment not in the multivariable analysis. Medical care in New Zealand is provided without cost to the patient. However, there are considerable indirect costs. Some are covered or partially covered by the healthcare system including travel and accommodation, while others—such as loss of income—may not be.

A recent study from the USA demonstrated that the maternal social vulnerability, an index of economic and social vulnerability similar to the NZ deprivation index, is associated with a delayed mid-trimester screening ultrasound and a longer interval to a fetal cardiology opinion in pregnancies affected by congenital heart disease.23 In the same study, maternal religious affiliation was also related to a delayed prenatal diagnosis, while in another US study of fetuses with transposition of the great arteries, rurality and Hispanic ethnicity were associated with a lower likelihood of prenatal diagnosis.24

Information that women receive can be interpreted differently depending on parental and family values, religious beliefs and language barriers. Communication with people from diverse ethnic, socioeconomic and health literacy backgrounds may be influenced by explicit and implicit bias.24,25 Implicit bias is an important contributor to healthcare disparity25-27 affecting decision-making, treatment recommendations, verbal and non-verbal communication and influencing the way in which diagnosis and treatment options are presented to different groups.25-27 There is evidence that ethnic disparities occur in the quality of healthcare delivery in New Zealand beyond inequities in access, with30 Māori more likely to report experiencing discrimination from a healthcare professional than other groups.27 Little is known about the experience of Māori families receiving a diagnosis and treatment plan for an infant with HLHS or the impact this experience has on prenatal or postnatal decision-making. Further studies are required to better understand Māori families’ experiences and to identify factors responsible for the observed inequity. This study spanned 10 years during which three fetal cardiologists and a larger number of MFM specialists were involved in the programme. Although the counselling process was standardised, the study was not designed to measure the impact of counselling on decision-making. We are currently investigating this issue prospectively with a mixed-methods approach in order to identify factors that influence decision-making.

The higher prevalence of later diagnosis among Māori and those living a distance from an MFM centre signal a need to address inherent system-related barriers to maternal care, develop culturally safe care systems and direct resources equitably. The current health system reforms, in particular the focus on equity, work on the ‘kahu taurima’ (maternity and early years) workstream, and the increasing focus on cultural safety for health professionals provide opportunities to realise these required changes. Functions of Te Aka Whai Ora—the newly formed Māori Health Authority—including ensuring that Māori are involved in the design and implementation of healthcare services and systems, and monitoring
service delivery and outcomes to ensure that inequities are identified and addressed.

Established surgical risk factors, including RV dysfunction and major non-cardiac abnormalities were associated with outcome in livebirths with an intention-to-treat. Surgery did not proceed in five infants in this group on the basis of previously unrecognised cardiac or non-cardiac comorbidity that became evident after birth. Case review indicated mutual agreement by the family and the treating surgeons and physicians in each of these instances. Nevertheless, all five of these infants had non-European mothers. These findings need to be interpreted with caution; the small sample size precludes multivariable analysis and differences in termination rates across ethnic groups may confound differences in postnatal outcome.

The major determinants of 30-day postoperative survival relate to the underlying cardiac condition and to associated comorbidities. We have previously found an association between outcome up to the second stage surgery (3–6 months of age) and socioeconomic position, and others have demonstrated an association between this outcome measure and poverty and ethnicity. It is likely access to healthcare and other resources becomes an important determinant of survival after hospital discharge.

This study is unique in that it captures a cohort that is near-complete from 20 weeks’ gestation. The concentration of fetal cardiology and cardiac surgery in one centre and the network nature of the MFM hubs results in uniformity of care pathways and provides a robust mechanism to track cases that is lacking in most other jurisdictions. Incomplete case ascertainment and case tracking is problematic, complicating the interpretation of findings and reducing their impact. The inclusive nature of the current dataset clearly illustrates the impact of access and inequity on the CCP and provides a means to assess the efficacy of targeted and system-wide interventions.

Conclusions

In this population-based cohort of pregnancies with a prenatal diagnosis of HLHS, the CCP was associated with gestational age at diagnosis, and care from the largest, most sparsely populated MFM region. Gestational age at diagnosis was in turn associated with ethnicity and distance from the MFM hub. Anatomic factors were not associated with CCP before birth but did influence the decision to treat after birth, and the outcome of cardiac surgery. These findings reinforce the importance of access to healthcare in determining the CCP. The association of ethnicity with delayed diagnosis before birth and decision-making after birth is indicative of systemic inequity and requires further investigation.

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