



Incidence and aetiology of talipes equino-varus with recent population changes

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This study looks at the changing incidence and aetiology of congenital talipes equinovarus due to the recent population changes within the area.

Between 1st June 1992 and the 31st May 2006, 83 consecutive children (121 feet) born with fixed talipes equinovarus (TEV) were assessed and treated (an incidence of 1.6 per 1000 live births) in an observational longitudinal cohort study assessing associated factors.

There were 17 syndromal cases in the fixed group (20.8%), 6 cases of non-syndromal distal arthrogyrosis (7.2%), and a strong family history in 12 cases (14.5%).

This study would suggest that genetic and primary causes of fixed TEV are more common than previously considered. Many of the primary aetiologies were diagnosed months or years after birth.

Keywords : talipes equinovarus ; incidence ; aetiology ; population changes.

This gives a more current representation for the local population that has evolved significantly in the last generation, principally due to immigration from the Asian subcontinent.

MATERIALS AND METHODS

Between 1st June 1992 to the 31st May 2006, all children with fixed talipes equinovarus (TEV), postural equinovarus and calcaneovalgus born in the Blackburn, Hyndburn and Ribble Valley District were referred to the senior author (RWP) and data was collected in a prospective manner. In non-idiopathic feet, the primary cause of the fixed talipes equinovarus (syndromes and neurological causes) were noted. Any syndromal diagnosis, or arthrogyrosis (including distal types), was made by the Paediatricians, Paediatric neurologists or Clinical geneticists. Cases referred with fixed TEV were assessed by

INTRODUCTION

A clubfoot (TEV) service has run in the Blackburn District since 1992. The Paediatric department refers all cases of fixed and postural talipes equinovarus to the senior author (RWP). All data on club feet, including idiopathic, acquired, neurogenic and postural, is documented prospectively and this data has been analysed to assess incidence and aetiology of this particular condition in a district with a significant immigrant population.

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the senior author within days of the birth. Foot deformities were classified using the Harrold & Walker classification (5), a simple method of clinical evaluation of the severity of fixed club foot deformity, validated by MacNicol, which correlates reasonably well with outcome (8). Additionally, all cases of fixed talipes equinovarus foot referred underwent ultrasound screening of the hips by the senior author soon after birth, as foot abnormalities are a risk factor for developmental dysplasia of the hip (DDH) (12). Postural clubfeet were seen for assessment of the feet and for ultrasound screening of the hips usually at 6-10 weeks of age.

A family history of fixed TEV in siblings, parents and first cousins was sought. The need to wean neonates off heroin secondary to maternal heroin addiction was recorded. Data from the Office of national Statistics (10), the Department for Communities and Local Government (formerly the Office of the Deputy Prime Minister until June 2006) (4), and the UK National census 1991 and 2001 (9), were used to assess the degree of deprivation and ethnicity statistics for the population.

All cases of fixed talipes equinovarus were initially treated by conservative methods : serial casting/dynamic splintage (1992-2002) or the Ponseti approach (2002-2006) (13).

RESULTS

The birth rate mean in the Blackburn, Hyndburn and Ribbles Valley Districts was 3,692 per year (51,693 over the 14 year period).

Over this period 83 individuals, 121 clubfeet, were referred with fixed talipes equinovarus. There were 61 males and 22 females, a male :female ratio of 3:1. There were 38 bilateral and 45 unilateral deformities, and 65 right and 56 left feet.

The incidence of all cases of fixed TEV was 1.6 per 1000 live births and excluding syndromal, arthrogryptic and drug abuse cases, the incidence of idiopathic TEV decreases to 1.1 per 1000.

There were 4 deaths secondary to the effects of severe syndromal disease. There were 17 cases (20.5%) with a primary syndromal aetiology (table I) and 6 cases (7.2%) of distal arthrogryposis. There was a strong family history in 12 neonates (14.5%), none of which had an associated syndrome. Overall, 27.7% of this group of fixed TEV (N = 83) had a primary syndromal or arthrogryptic underlying aetiology with a further 14.5% having

Table I. — Syndromal causes

1. Microcephaly/developmental delay
2. Ehler-Danlos syndrome
3. Syndactyly hands & feet, hypoplasia of the toes & epilepsy
4. Generalised myopathy (died)
5. Unknown syndrome with multiple abnormalities (died)
6. Bilateral dislocated hips with fractured femur in neonatal period, unknown syndrome (died)
7. Metabolic syndrome
8. Klinefelter's syndrome with scoliosis & hydronephrosis
9. Short stature with developmental delay (syndrome unknown)
10. Cleft palate and arthrogryposis
11. Absent little & ring fingers & metacarpals, severe leg length discrepancy (on side of TEV)
12. Di George syndrome (VSD aortic arch incomplete)
13. Severe developmental delay (unknown syndrome)
14. Very short stature (unknown syndrome)
15. Fallot's tetralogy with severe learning difficulties (died)
16. Myelomeningocele
17. Myelomeningocele

an unidentified genetic predisposition. There were 29 (34.9%) individuals from the Asian community. Eight of the 17 syndromal (47%) and 5 of the 12 cases with a strong family history (41.6%) were from the Asian community. In our group of fixed TEV, occurrence of a syndrome amounted to 27.6% in Asian individuals (n = 29), compared to 16.7% in the Caucasians. Incidence of family history was similar between the Asian and Caucasian neonates (17.2% and 13.0% respectively).

Four individuals were born to known heroin abusers and all these children underwent weaning off the drug in the neonatal period (all 4 cases were caucasian).

All the fixed clubfeet (apart from the 2 cases of myelomeningocele, and one case of Fallots' tetralogy, which were not referred at birth) were assessed by the Harrold and Walker classification at the first treatment session. There were 56 (47.9%) grade 3, 39 (33.3%) grade 2 and 22 (18.8%) grade 1 deformities.

All individuals underwent hip ultrasound screening ; no cases of dislocation nor developmental dysplasia of the hip were identified, and we believe that talipes equino-varus alone is not a risk factor

for hip dysplasia, which is supported in the literature (11,17).

In addition to the 83 fixed talipes equinovarus cases, there were a further 424 cases of postural TEV, none of which required treatment. This equates to a ratio of postural to fixed TEV of just over 5:1.

DISCUSSION

Wynn Davies noted that talipes equinovarus has a multifactorial inheritance pattern with both environmental and genetic causes, with a ratio of male to female equalling 2:1 (18). Talipes equinovarus can have an idiopathic or acquired cause (1,7) and the incidence of congenital talipes equinovarus worldwide varies from 0.64 to 6.8 per 1000 live births (1). There are strong racial and genetic influences in different parts of the world (7,18). In the United Kingdom the rate of CTEV is approximately 1.24 per 1000 (18) and our study is broadly in line with this, at 1.6 per 1000. It is generally agreed in the UK, that there is a polygenetic inheritance for sensitivity to unknown environmental factors predisposing to CTEV (18).

In this series, a strong family history was present in 14.5% of cases, which is slightly greater than another equivalent UK series with 11.6% (5), though due to the small sample size, this difference may not be statistically valid.

Seasonal variation has been observed in several studies (15,18), though not in others (6) and again no variation could be found in this series. Various other factors have been linked to congenital talipes equinovarus including maternal smoking and alcohol consumption (1), however this is difficult to substantiate and quantify and we do not have data on this.

Non idiopathic/acquired causes associated with syndromal cases of TEV include spina bifida, myelomeningocele, sacral agenesis, arthrogryposis, congenital myopathy, constriction-band syndrome, Freeman-Sheldon syndrome (distal arthrogryposis), foetal alcohol syndrome and Down's syndrome (1,7). The diagnosis of a syndromal cause cannot always be made at birth and this can lead to an under reporting of these particular cases. Many

of our syndromal cases were diagnosed months or years after birth. This study would suggest that severe, bilateral or resistant talipes equinovarus cases should be investigated for underlying primary aetiology and continued surveillance of the child is appropriate.

As the ratio of postural to fixed TEV was greater than 5:1 in this study, the importance of recording the initial foot deformity in reporting the results of treatment is obvious. If accurate pre-operative assessment is not undertaken, inclusion of postural TEV may bias the results, (a Type 1 statistical alpha error) (14). However, the diagnostic criteria separating postural (or positional) TEV from the fixed type are not well defined in the literature. If full dorsiflexion of the foot is not achieved with greater than 20° of dorsiflexion from plantigrade, this is technically a fixed deformity. The Stevens and Meyer classification (16), is useful in differentiating between true postural and mild fixed cases. In the Harrold and Walker study there were 37.9% grade 1, 24.8% grade 2, and 37.2% grade 3, compared with 18.8% grade 1, 33.3% grade 2, and 47.9% grade 3, in our study. It is possible that the low number of grade 1 types in our study is due to the exclusion of all postural TEV. The number of grade 3 feet may reflect the relatively high proportion of primary causes highlighted in this study.

To explain this significant incidence of underlying etiologies, the environment and possible genetic differences were investigated in an attempt to explain this. The district includes Blackburn with Darwin (population 140,000) with a Standard Mortality Ratio (SMR) of 124, Hyndburn (population 81,000) with an SMR 113, and the Ribble Valley (population 61,000) with an SMR of 98 (10). The average SMR for England and Wales is 100. A SMR of 124 indicates that there are 24% more deaths than expected if it had the same age and sex mortality rates of England and Wales. Assessing the Indices of Deprivation 2004 (ID 2004) (4), Blackburn and Darwin are in the bottom 10% and Hyndburn is in the bottom 20% of the most socially deprived districts in England. The Ribble Valley is in the top 2-14% of least deprived districts in England. This indicates that the majority of this study's population (around 80%) are among the

most deprived groups in England and this may be an important environmental factor in the development of TEV.

The level of intravenous drug abuse is higher than average ; the Northwest region has the highest level of drug abuse in England and Wales (out of 10 regions) in those age 16 to 59 years (3). The use of class A drugs (which includes heroin and cocaine), in the Northwest region is recorded as the third highest level of the 10 regions. In our series, there were four cases in which the neonate had to be weaned off heroin at birth. All these feet were very rigid and severe (Harrold and Walker grade 3) requiring extensive surgical release. The effects of substance abuse on the developing limbs in the foetus during early pregnancy is unknown, and although there is some evidence that substance abuse may be teratogenic (1), this subject requires further research.

The Blackburn and Darwin districts, have the second highest birth rate in England. The average percentage of Blackburn Asian births was 26% over the past 16 years according to our pediatric departments database. There is a significant degree of consanguinity within the Asian population. In some UK districts, 60% of the marriages in the Pakistani and Guterati population were thought to be consanguineous. The Asian population in Blackburn has a 12 fold increased risk of recessive disorders compared with the caucasian population (2). Consanguinity is thought to be a major contributor to this. This may account for some of the cases of syndromal disease, though as idiopathic TEV is polygenetic, the autosomal recessive gene should not be involved in these cases (16). As 34.9% of the total of fixed TEV was in the Asian population, genetic factors may be more important in the Asian compared with Caucasian population in the development of TEV in our district.

In conclusion, this study has shown a relatively high percentage of fixed Congenital Talipes Equinovarus presenting with an acquired cause such as arthrogryposis, a positive family history, a syndrome, or possibly secondary to maternal drug abuse. Acquired causes may be more important than previously considered in the aetiology of TEV.

There is a need for further research into the incidence and aetiology of this condition, as the changing ethnicity of the UK population may change the accepted incidence and aetiology compared with previous studies in the 1970's (18).

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