The background

No specific standards exist for the management of Congenital Diaphragmatic Hernia (CDH), although guidelines for neonatal management are readily available. Two Cochrane reviews pertain to the management of CDH (iNO and late vs early surgery).

The generic Standards for MCN include the following

Standard 1: Organisation – Management Arrangements and Accountability
Standard 2: Clinical – Delivering CDH clinical network care
Standard 3: Multidisciplinary working
Standard 4: Staff education and Training
Standard 5: Audit, Monitoring, Research and Development
Standard 6: Patient Focus and Public Partnership

Most other MCN have developed at least 5-6 standards.

Individually developed standards should:

- have a sound evidence base
- focus on clinical issues
- directly relate to the objectives of the MCN
- be clear and measurable
- follow the patient pathway, and
- be consistent with other MCNs of the same topic across Scotland

From the generic standards, I suggest that we should have at least one standard from the clinical subgroups (generic standard 2), and one standard from each of the categories 3-6.

Suggested standards for SDHCN:

Standard 1 Clinical – Antenatal Care
Standard 2 Clinical – Counselling
Standard 3 Clinical – Inpatient management
Standard 4 Clinical – Follow-up
Standard 5 Multidisciplinary working
Standard 6 Staff Education
Standard 7 Audit & monitoring
Standard 8 Patient information
Standard 1: Clinical (Antenatal Care)

Standard Statement 1:
Following identification of a congenital diaphragmatic hernia (CDH) on booking scan or fetal anomaly scan (FAS), the expectant mother will be offered a further detailed examination at a specialist fetal medicine service within 2-5 working days.

Rationale
At present, the best available prognostic antenatal criteria are based on liver position, lung head ratio and fetal echocardiography.
Time interval to referral is based Fetal Anomaly Screening Programme (FASP) guidelines.

Reference:
- [http://fetalanomaly.screening.nhs.uk/](http://fetalanomaly.screening.nhs.uk/)

Essential Criteria

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1.1</td>
<td>With the introduction of routine anomaly scans, the expectation is that around 60% of patients will be identified antenatally.</td>
</tr>
<tr>
<td>1.2</td>
<td>A minimum dataset for antenatal scans has been agreed by the SDHMCN.</td>
</tr>
<tr>
<td>1.3</td>
<td>Data on fetal anatomical and physiological parameters are collated in a location that is accessible for staff involved in counselling, perinatal and postnatal treatment.</td>
</tr>
</tbody>
</table>

Indicator of Standard

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Referred to Specialist Fetal Medicine Unit (Y/N)</td>
</tr>
</tbody>
</table>
Standard 2: Clinical (Antenatal Counselling)

Standard Statement 2:
When a CDH has been diagnosed antenatally, families will be offered multidisciplinary counselling. Ideally, counselling should include obstetricians, neonatologists, and neonatal surgeons but not necessarily all at one visit.

Rationale
To ensure that families are fully informed of the likely peri- and postnatal management possibilities.

Reference:

Essential Criteria

2.1 An information leaflet that has been designed and endorsed through the SDHMCN will be offered to the families in conjunction with counselling.

2.2 Counselling should ideally be planned to occur during two separate sessions in the second and third trimester.

Indicator of Standard

2 Counselled by Obstetrician/Neonatologist/Surgeon (Name, Date)
Standard 3: Clinical (Inpatient Management)

<table>
<thead>
<tr>
<th>Standard Statement 3:</th>
<th>Neonates born outwith specialist centres should be discussed with the relevant specialist centre within 2 hours of diagnosis.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rationale</td>
<td>To ensure that recommended management principles are employed from outset and to encourage discussion regarding optimal timing of transfer</td>
</tr>
<tr>
<td>Reference:</td>
<td>o SDHCN Steering Group opinion of best practice.</td>
</tr>
<tr>
<td>Essential Criteria</td>
<td>3.1 Each specialist centre should have a designated pathway for contact and discussion</td>
</tr>
<tr>
<td></td>
<td>3.2 The Neonatal Transport Service should be contacted by the referring hospital following discussion of the eligibility and appropriateness of transfer</td>
</tr>
<tr>
<td>Indicator of Standard</td>
<td>3 Time to Referral (measured in “Birth Details”)</td>
</tr>
</tbody>
</table>
Standard 4: Clinical (Inpatient Management)

Standard Statement 4:
Newborns with CDH will be managed according to agreed guidelines that have been ratified by the SDHCN

Rationale
Survival from CDH has been shown to improve with adoption of neonatal management guidelines.

Reference:

Essential Criteria

| 4.1 | Inpatient management guidelines, produced by the SDHMN, offer guiding principles for management of ventilation, manipulation of pulmonary vascular reactivity and timing of surgery |
| 4.2 | Principles of management will be available on the website |
| 4.3 | Aspects of management will be discussed at educational study days to ensure information is current |

Indicators of Standard

| 4a | Repair done (Y/N) |
| 4b | If no repair done, reason why not |
| 4c | Maximum Peak Inspiratory Pressure on IMV |
| 4d | Highest and lowest pCO₂ in first 24 hours |
### Standard 5: Multidisciplinary Working & Follow-up

#### Standard Statement 5:
Outpatient follow-up will be conducted to an agreed protocol

#### Rationale
Children who survive CDH are known to have long-term morbidity, with some aspects only becoming apparent at a later stage

#### Reference:

#### Essential Criteria

| 5.1 | Specific needs should be assessed for individual patients with emphasis on nutritional, respiratory and developmental outcome |
| 5.2 | Multiple health care practitioners may be involved in ongoing care and these personnel should be aware of the potential morbidity in CDH |
| 5.3 | Local follow-up should be encouraged if adequate resources are available to offer a streamlined service |

#### Indicator of Standard

| 5a | Record of different specialities that have reviewed in out-patients |
| 5b | Post-discharge Hearing screening |
Standard 6: Staff Education

Standard Statement 6:
The Network will provide regular education for staff involved in managing patients with CDH and each specialist centre will be represented and will encourage and facilitate staff to attend. Telehealth links will be utilised to facilitate this where appropriate.

Rationale
Ensure staff are aware of current management options and outcome data

Reference:
- [http://www.sdhcn.nhs.uk](http://www.sdhcn.nhs.uk)
- [http://www.sctt.nhs.uk/](http://www.sctt.nhs.uk/)

Essential Criteria

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>6.1</td>
<td>The Network will arrange yearly educational days with locations to be rotated to encourage attendance from all centres</td>
</tr>
<tr>
<td>6.2</td>
<td>Topics covered will include presentations that will focus on each discipline involved in the care of families and patients with CDH</td>
</tr>
<tr>
<td>6.3</td>
<td>Educational days will be advertised in all paediatric and maternity units with adequate notice</td>
</tr>
</tbody>
</table>

Indicator of Standard

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>Network to record date of education day, topics and attendance (each centre)</td>
</tr>
</tbody>
</table>
Standard 7: Audit & Monitoring

Standard Statement 7:
The network will record activity, management and outcome of patients with CDH

Rationale
A national database will provide a greater understanding of the aetiology, demographics, management strategies and outcome for CDH in Scotland.

Reference:
- Doyle NM, Lally KP. The CDH Study Group and advances in the clinical care of the patient with congenital diaphragmatic hernia. Seminars in Perinatology;28(3):174-84, 2004

Essential Criteria

| 7.1 | Completion of SDHCN core dataset for CDH by each centre |
| 7.2 | Regular distribution of data during staff education days, website and newsletters |

Indicator of Standard

| 7 | Network completion rate of core dataset |
Standard 8: Patient Information

Standard Statement 8:
An information leaflet will be distributed to parents of children with CDH (including prospective parents) with details of possible clinical expectations and links/contact details for further information and support.

Rationale
Parents will be provided with an information leaflet that covers antenatal and post-natal management possibilities to reinforce/augment information given at counselling, during in-patient stay and following discharge (or death).

Reference:

Essential Criteria

8.1 SDHCN has produced written information covering antenatal and postnatal management

8.2 The leaflet is also be available as a document on the website

8.3 The leaflet will be reviewed every 2 years to ensure accurate contemporaneous information is included

Indicator of Standard

8a Parents offered Information leaflet antenatally (in “Birth details”)

8b Parents offered Information leaflet after birth (in “Outcome-Discharge”)
Summary of Standard Statements

Standard Statement 1:
Following identification of a congenital diaphragmatic hernia (CDH) on booking scan or fetal anomaly scan (FAS), the expectant mother will be offered a further detailed examination at a specialist fetal medicine service within 2-5 working days.

Standard Statement 2:
When a CDH has been diagnosed antenatally, families will be offered multidisciplinary counselling. Ideally, counselling should include obstetricians, neonatologists, and neonatal surgeons but not necessarily all at one visit.

Standard Statement 3:
Neonates born outwith specialist centres should be discussed with the relevant specialist centre within 2 hours of diagnosis.

Standard Statement 4:
Newborns with CDH will be managed according to agreed guidelines that have been ratified by the SDHMCN.

Standard Statement 5:
Outpatient follow-up will be conducted to an agreed protocol.

Standard Statement 6:
The Network will provide regular education for staff involved in managing patients with CDH and each specialist centre will be represented and will encourage and facilitate staff to attend. Telehealth links will be utilised to facilitate this where appropriate.

Standard Statement 7:
The network will record activity, management and outcome of patients with CDH.

Standard Statement 8:
An information leaflet will be distributed to parents of children with CDH (including prospective parents) with details of possible clinical expectations and links/contact details for further information and support.