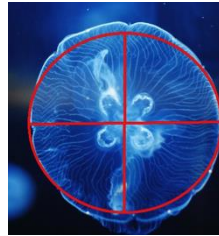# Coding Defects of Bilateral Symmetry

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Symmetry

Radial: Arrangements of parts around a central axis such that the object can be divided into similar halves by any plane containing that axis.
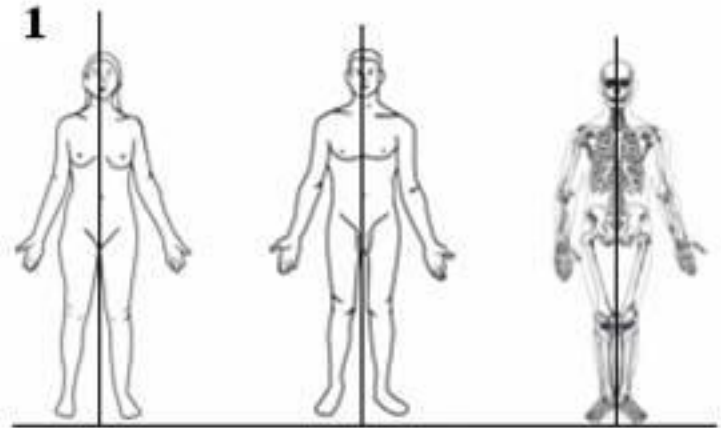

Bilateral: Arrangement of parts so that division along the central axis creates two halves that mirror each other.
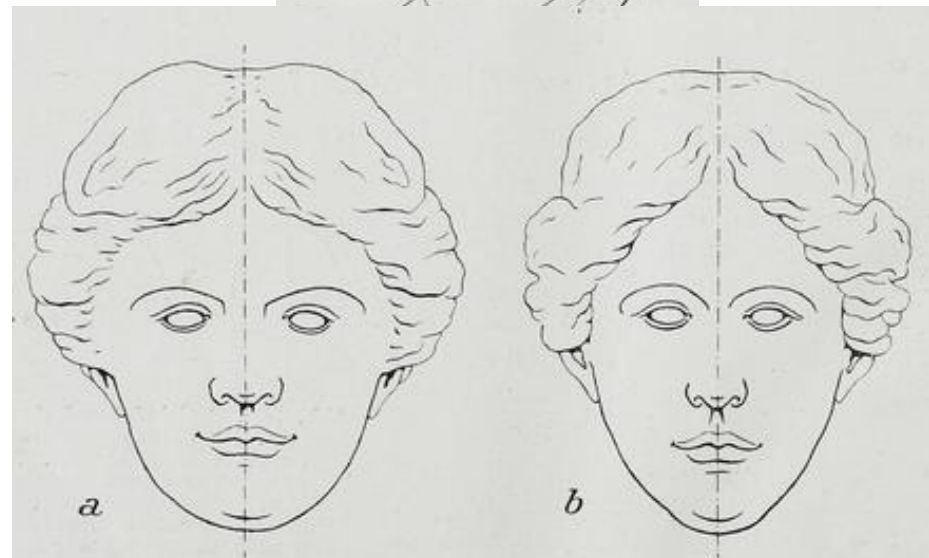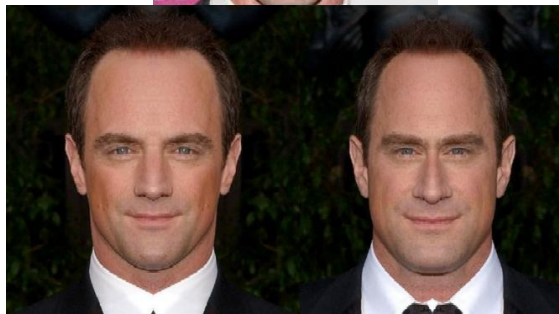

© 1992 Jonathan Bird

Humans

- Are (mostly) bilaterally symmetric
- Big exceptions
  - Great vessel connections, Lung lobation, GI tract starting at the stomach, spleen, liver and gall bladder

Humans

- Also have little asymmetries
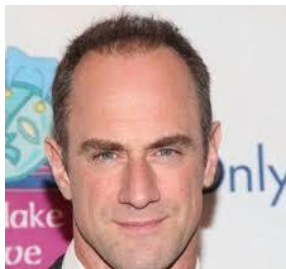
We have to decide

- Normal Asymmetry
  - Left handed
  - Nearsighted in right eye
  - Right ventricle more prominent
  - Right kidney lower
  - One testis larger

- Abnormal Asymmetry
  - Missing phalanx right 5th finger
  - Left esotropia
  - Enlarged right ventricle
  - Ectopic right kidney
  - Absent left testis

Coding Asymmetry (IMO)

- Simple asymmetry itself is not a birth defect.
- There must be a clearly abnormal side.
- The abnormal side may be the one that is too big, or it may be the one that is too small
- We can't assume that the side that is mentioned is the abnormal one:
  - "Right [organ] is larger than left [organ]" doesn't tell us which is abnormal

Arbitrary Agenda

- "Isolated" defects and syndromes
  - Orofacial clefting
  - Facial palsy
- Conditions that imply/require other codes
  - Heterotaxy
  - Hemihypertrophy/Hemihyperplasia
  - Hemifacial microsomia
  - Poland Anomaly
- Multiple malformation syndromes
  - Moebius Syndrome
  - Russell-Silver Syndrome
  - CHILD Syndrome
  - Proteus Syndrome

Single Codes

- Cleft lip

- Cleft lip with cleft palate

- Asymmetric Crying Facies/Facial Palsy

Cleft Lip

Defect	BPA	ICD 9	ICD 10
Cleft lip NOS	749.190	749.10	Q36.9
Unilateral Complete cleft lip	749.100*	749.11	Q36.9
Unilateral Incomplete cleft lip	749.100	749.12	Q36.9
Bilateral Complete cleft lip	749.110	749.13	Q36.0
Bilateral Incomplete cleft lip	749.110	749.14	Q36.0
Central cleft lip	749.120		Q36.1

The codes do not provide for coding right/left laterality.
Laterality will need to be indicated another way.

*Texas uses this code for any cleft other than central, then indicates the laterality as a separate data field.

Cleft Lip with Cleft Palate

Defect	BPA	ICD 9	ICD 10
Cleft lip NOS with any cleft palate	749.290	749.20	Q37.9
Unilateral Complete cleft lip with cleft palate	749.200*	749.21	Q37.5
Unilateral Incomplete cleft lip with cleft palate	749.200	749.22	Q37.9
Unilateral lip/Hard palate			Q37.1
Unilateral lip/Soft palate			Q37.3
Unilateral lip/Hard+Soft			Q37.5
Unilateral lip/Unspecified palate			Q37.9

The codes do not provide for coding right/left laterality.
Laterality will need to be indicated another way.

*Texas uses this code for any cleft other
than central, then indicates the
laterality as a separate data field.

Cleft Lip with Cleft Palate

Defect	BPA	ICD 9	ICD 10
Bilateral Complete cleft lip with cleft palate	749.210	749.23	Q37.4
Bilateral Incomplete cleft lip with cleft palate	749.210	749.24	Q37.8
Other combinations		749.25	Q37.9
Central cleft lip with any cleft palate	749.220		NONE!?!
Bilateral lip/Hard palate			Q37.0
Bilateral lip/Soft palate			Q37.1
Bilateral lip/Hard+Soft			Q37.4
Bilateral lip/Unspecified palate			Q37.8

Facial Palsy

What gets coded depends on what is charted.

Defect	BPA	ICD 9	ICD 10
Asymmetric Crying Facies	351.000	351.0	G51.0
Facial Palsy (Weakness)		781.94	
Hypoplastic depressor angularis	744.880	756.81?	Q79.9?

The codes do not provide for coding right/left laterality.
Laterality will need to be indicated another way.

Other Codes Required

- Heterotaxy

- Hemihypertrophy/Hemihyperplasia

- Hemifacial microsomia

- Poland Anomaly

Heterotaxy

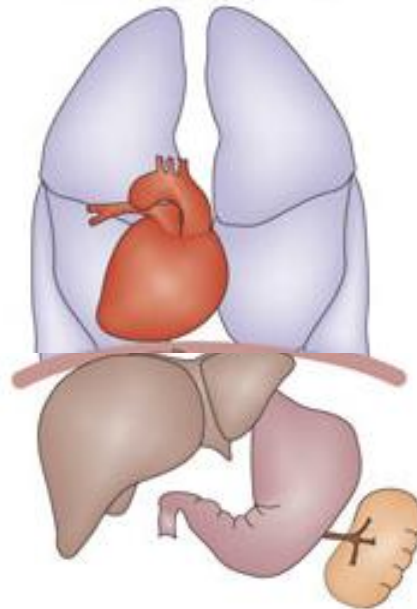| Defect	BPA	ICD 9	ICD 10
Complete Situs Inversus	759.300	759.3	Q89.3
Situs Inversus with Levocardia	759.310	759.3	Q89.3
Situs Inversus Thoracic Reversal of Thorax/Abdomen normal	759.320	759.3	Q89.3
Situs Inversus Abdominis Reversal of Abdomen/Thorax normal	759.330	759.3	Q89.3
Ciliary Dyskinesia* Kartagener syndrome	759.340	759.3	Q89.3

Heterotaxy NOS*
    Abnormal Thorax and/or Abdomen NOT
    mirror image reversal
	759.390	759.3	Q89.3

*Require coding of the individual organ abnormalities, because they cannot be assumed.

Heterotaxy

Normal

Lungs

Heart

Liver

Spleen

Stomach
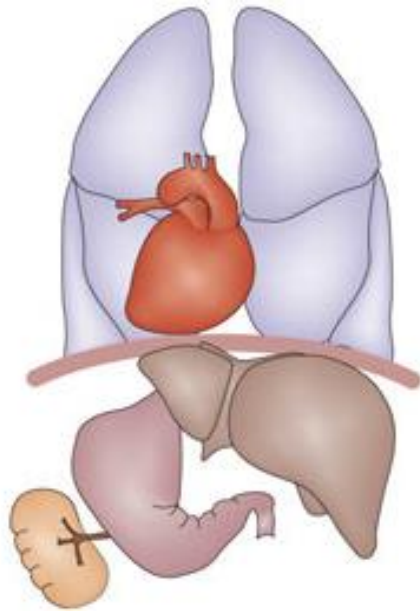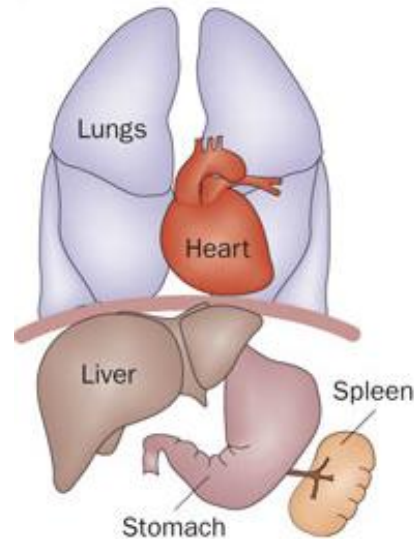
Complete Situs Inversus
759.300

Situs Inversus Thoracis
759.320
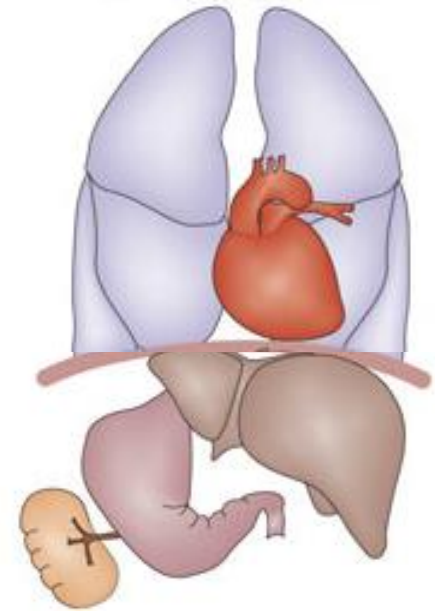
Situs Inversus Abdominis
759.330

"Situs"

Situs Solitus

Anomalías del Situs

Situs Inversus

Situs Ambiguous

Heterotaxy NOS
759.390

Dextrocardia

Levocardia

Asplenia

Poliesplenia *

Complete Situs
Inversus
759.300

Situs
Inversus w
Levocardia
759.310

Right
Isomerism
759.390

Left
Isomerism
759.390

Adapted from
Rev Chil Radiol 2013;19(1)
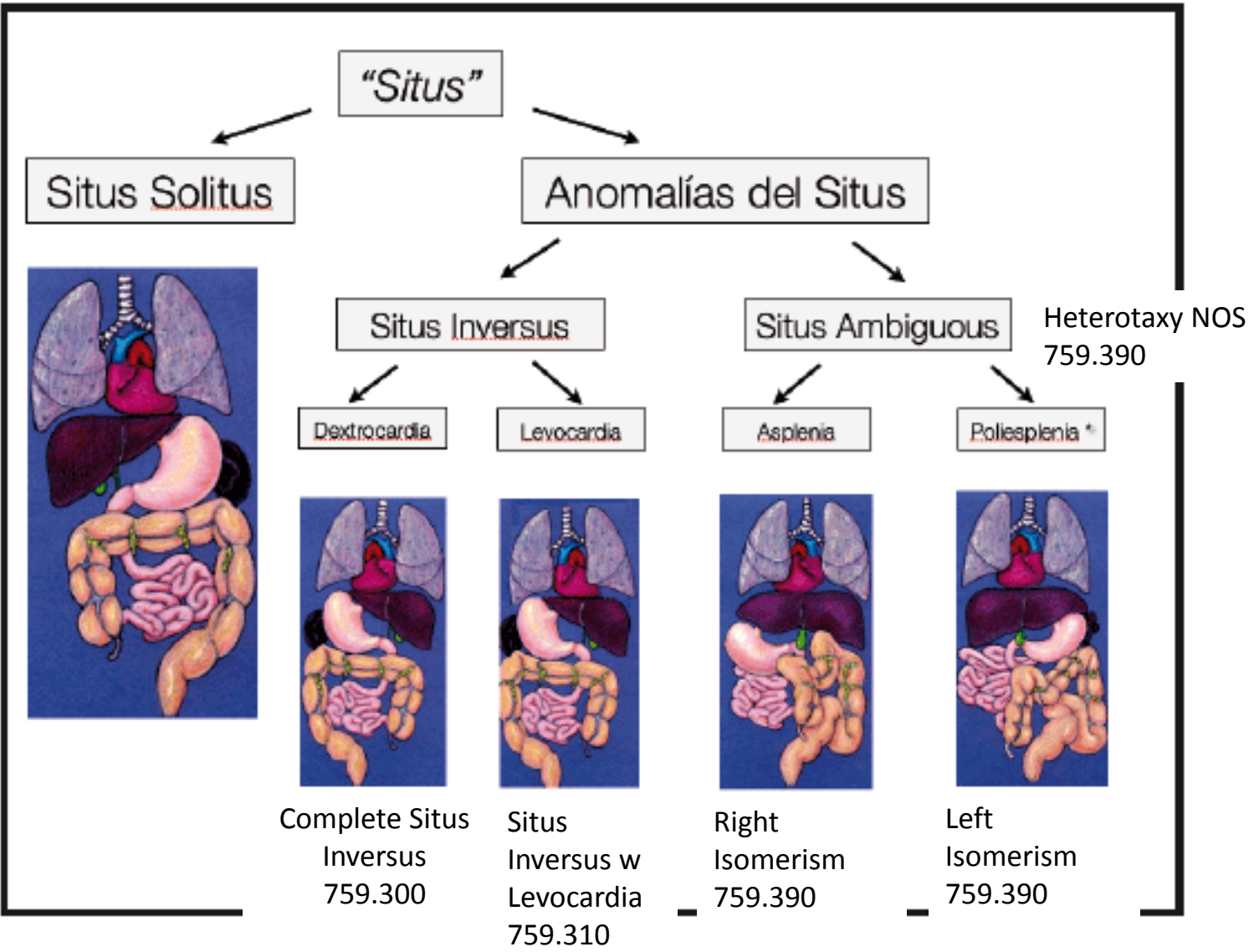
Heterotaxy

- Some defects limited to the heart are called heterotaxy.
  - Recommendation: code as the heart defect, not as a situs abnormality
- "Asplenia" and "Polysplenia" are used as nicknames
  - Don't rely upon them as a good description of the situs abnormality
- Kartagener is mistakenly used as
  - A nickname for situs abnormalities
  - Indicative of a specific single gene disorder

Heterotaxy – Specific Organs

Defect	BPA	ICD 9	ICD 10
Dextrocardia	746.800	746.87	Q24.0
Various congenital heart defects	745,xxx- 747,xxx	745.xx- 747.xx	Q20 – Q28
Malpositioned liver	751.620	751.69	Q44.7
Malpositioned/absent/extra spleen	759.0xx	759.0	Q89.0
Intestinal malrotation	751.400 or 751.490	751.4	Q43.3

When coding Heterotaxy with 759.340 or 759.390, code the other reported organ abnormalities as separate items.

Hemihypertrophy/Hemihyperplasia

- Part(s) of the body are overgrown
- The LARGE side is the abnormal one
  - Limb
  - Half of face
  - Part of the thorax
- Found as an isolated defect or in some syndromes
  - Beckwith-Wiedemann Syndrome

Hemihypertrophy/Hemihyperplasia

Hemihypertrophy/Hemihyperplasia

Defect	BPA	ICD 9	ICD 10
Hemihypertrophy/Hemihyperplasia	759.890	759.89	Q87.8

But then the specifics are necessary:

Defect	BPA	ICD 9	ICD 10
Musculoskeletal abnormalities (General)	756.880	754.89	Q18.8
Hemifacial Hypertrophy	744.880	744.89 or 754.0	Q67.4
Abn chest	754.820	754.89	Q67.8
Abn upper limbs	755.5XX	755.59	Q74.0
Abn lower limbs	755.6XX	755.69	Q74.2

Hemifacial Microsomia

- Facial asymmetry in which the SMALL side is abnormal

- Broad spectrum condition

- Non-specific term used differently by different specialties

- "Isolated" finding

- May be associated with problems below the neck

OAV

OculoAuriculoVertebral Spectrum
(OAV)

Microtia ←——————————————————————→ Goldenhar

"Goldenhar" gets used as a nickname

Hemifacial Microsomia

Defect	BPA	ICD 9	ICD 10
Hemifacial Microsomia	756.065	754.0	Q67.0

But then the specifics are necessary:

Defect	BPA	ICD 9	ICD 10
Microtia	744.210	744.230	Q17.2
Asymmetric jaw	524.000	526.89 or 754.0	M26.12
Ear/facial tags	744.110	744.1	Q17.0
Lateral mouth cleft	744.800	744.83	Q18.4
Microtia	744.210	744.23	Q17.2

Poland Anomaly

- Unlateral absence of the pectoralis major muscle

- May be associated with hypoplasia of the ipsilateral arm

- (There's also a gluteal/leg version)

- It's most appropriately "Anomaly"
  - It's not a syndrome

Poland Anomaly
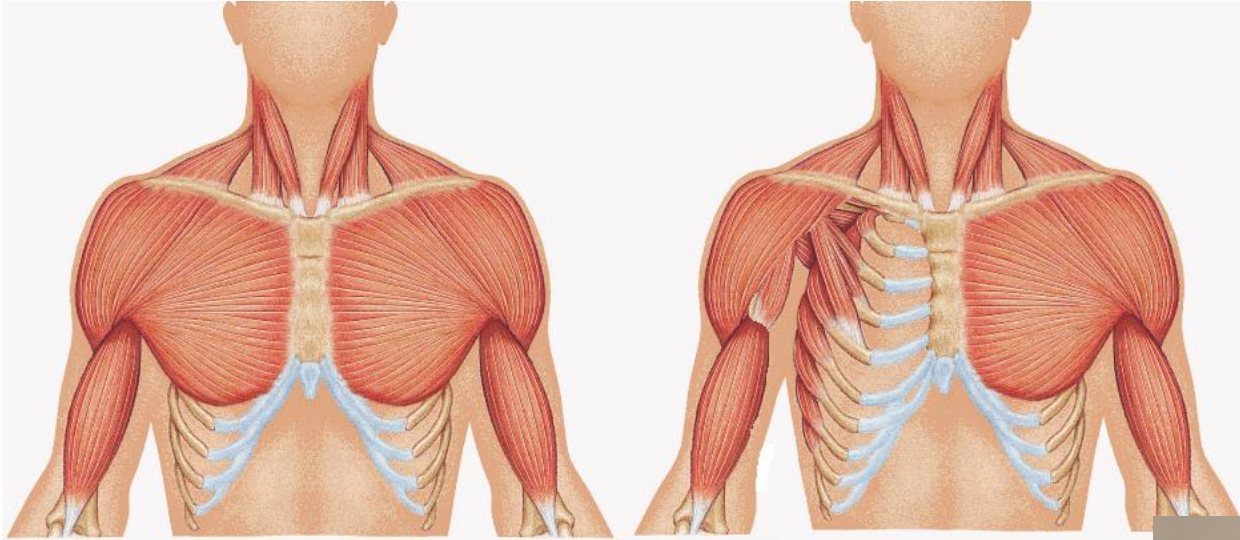
Typical symmetry seen in
pectoralis major muscles

Asymmetry seen due to
absent pectoral muscle

Poland Anomaly

Defect	BPA	ICD 9	ICD 10
Poland Anomaly	756.800	756.81	Q79.8

But then the specifics are necessary:

Defect	BPA	ICD 9	ICD 10
Absent Pectus muscle	756.810	756.81	Q79.8
Absent breast +/- absent nipple	757.600	757.6	Q83.0
Absent nipple alone			Q83.2
Hypoplasia of breast and nipple	757.610	611.82	N64.82
Hypoplasia nipple	757.640		Q83.8
Hypoplastic arm/hand (no reduction defect)	755.5xx	755.5x	Q74.0
Reduction defects of upper limb	755.2xx	755.2x	Q71.xx

Multiple Malformation Syndromes

- Moebius Syndrome

- Russell-Silver Syndrome

- CHILD Syndrome

- Proteus Syndrome

- All previous information applies.

- For all of these, the individual defects should be coded.

Moebius (Mœbius, Möbius)

- Facial nerve palsy

- Originally classified as just the facial palsy

- Now is any non-syndromic collection of birth defects that includes a facial palsy

  - You may find any birth defect in a baby reported to have Moebius

Moebius

Defect	BPA	ICD 9	ICD 10
Moebius	352.600	352.6	Q87.0

Defect	BPA	ICD 9	ICD 10
Asymmetric Crying Facies	351.000	351.0	G51.0
Facial Palsy (Weakness)		781.94	
Hypoplastic depressor angularis	744.880	756.81?	Q79.9?

And then code all the other defects mentioned

Russell-Silver Syndrome

- (Or Silver-Russell Syndrome to the Europeans)
- Prenatal and postnatal growth deficiency
- Proportionate short stature with normal head
  - So head looks big
- Typical facial features, minor malformations
- May have body asymmetry with diminished growth of the affected side
  - HemiHYPOtrophy
  - The SMALL side is the abnormal side

Russell-Silver

Defect	BPA	ICD 9	ICD 10
Russell-Silver	759.820	759.89	Q87.1

759.820 is Congenital Malformation Syndromes Associated with Short Stature

Defect	BPA	ICD 9	ICD 10
Hypoplastic arm/hand (no reduction defect)	755.5xx	755.5x	Q74.0
Hypoplastic leg/foot (no reduction defect)	755.6xx	755.6x	Q74.2

CHILD Syndrome

- **C**ongenital **H**emidysplasia with **I**chthyosiform **E**rythroderma and **L**imb **D**efects
- X-linked dominant male lethal
  - Will see the full blown syndrome only in females
  - There are less severe phenotypes
- Part/half/sections of the body will have abnormal growth, skin abnormalities, and reduction defects

CHILD

Congenital **H**emidysplasia with **I**chthyosiform **E**rythroderma and **L**imb **D**efects

CHILD

Defect	BPA	ICD 9	ICD 10
CHILD	759.840 or 757.300	759.89	Q87.8

759.840 is Congenital Malformation Syndromes Involving Limbs

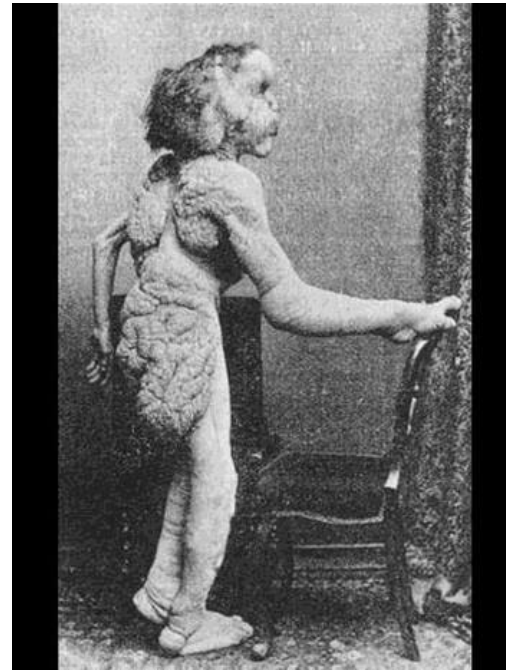| Defect	BPA	ICD 9	ICD 10
Reduction defect upper limb	755.2xx	755.2x	Q71.xx
Reduction defect lower limb	755.3xx	755.3x	Q72.xx
Ichthyosis	757.190 or 757.196	757.1	Q80.8 or Q80.1
Any associated internal malformation			

757.196 and Q80.1 = X-linked ichthyosis, which is usually going to be steroid sulfatase deficiency, but technically the code applies here.

Proteus

- Named after the Ancient Greek God who could appear in many different forms

- Progressive overgrowth of all tissues, mostly the skeleton, skin, fat and CNS.

- Lots of potential problems

- Any part of the body can be involved.
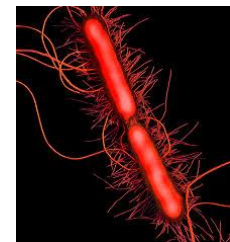
Proteus

Joseph Merrick

Proteus

Defect	BPA	ICD 9	ICD 10
Proteus	759.890 or 759.840	759.89	Q87.8

Be careful.
There is also a bacteria by this name.
It is common in hospital- acquired urinary tract infections.
It will show up in both ICD 9 and ICD 10 if you search on "Proteus".

Take Home Points

- "Isolated" defects and syndromes
  - Orofacial clefting
  - Facial palsy
- Conditions that imply/require other codes
  - Heterotaxy
  - Hemihypertrophy/Hemihyperplasia
  - Hemifacial microsomia
  - Poland Anomaly
- Multiple malformation syndromes
  - Moebius Syndrome
  - Russell-Silver Syndrome
  - CHILD Syndrome
  - Proteus Syndrome